PRIMARY TUMORS OF THE CALVARIA

With Special Consideration of the Clinical Problems

By

FRANKLIN JELSMA M D

Attending Neurosurgeon (Chief of Section) St Joseph Infirmary
Consulting Neurosurgeon Kentucky Baptist Hospital
Consulting Neurosurgeon St Anthony Hospital
Consulting Neurosurgeon Kosair Crippled Children Hospital
Assistant Professor of Neurosurgery University of Louisville School of Medicine
Louisville Kentucky



CHARLES C THOMAS • PUBLISHER
Springfield Illinois U.S.A

CHARLES C THOMAS PUBLISHER
BANNERSTONE HOUSE
301 327 East Lawrence Avenue, Springfield Illinois, U.S.A

Published simultaneously in the British Commonwealth of Nations by
BLACKWELL SCIENTIFIC PUBLICATIONS, LTD OXFORD ENGLAND

Published simultaneously in Canada by
THE RYERSON PRESS, TORONTO

Library of Congress Catalog Card Number 59-8498

Printed in the United States of America

TO MY WIFE, LILLIAN

PREFACE

The literature is decidedly limited on the subject of Primary Tumors of the Calvaria Except for 2 or 3 series of cases, one usually finds case reports, and not very many of these. Information concerning these rather rare lesions must be obtained in a piecemeal manner by searching through various text books dealing with bone tumors roentgenology pathology and widely scattered articles in the literature There is a need for clinical consideration of the subject as a whole. So it is thought worth while to place under one cover the various clinical aspects of these tumors so that this information will be more readily available

This monograph contains a classification of calvarial tumors, together with their symptoms diagnosis and treatment Operative and nonoperative viewpoints are discussed A brief discussion of the roentgenological findings and pertinent laboratory data is given Many case reports with illustrations are included

The arrangement, as planned in the classification of tumors in the first chapter has been carried out in order of sequence throughout the entire monograph The benign tumors are taken up first in the same order as listed in the table, the tumor like lesions are second in order and the malignant tumors are third This applies to the synopsis of cases as well as the arrangement of material case reports and illustrations throughout the entire work

Metastatic tumors of the calvaria, or involvement in a secondary manner, such as, in meningiomas are not mentioned except for reasons of differential diagnosis Tumors of the base of the skull such as, ethmoid groove osteomas or epidermoid sinus tracts extending to intracranial epidermoid cysts, are not included Encephaloceles are not considered primary skull tumors

The material comes from the Author's case records covering a period of over twenty years and includes our experiences in dealing with more than forty nine cases of primary tumors of the calvaria Only four cases of hyperostosis frontalis interna have

been included in this study Many, many more cases are present in our case records Two cases of xanthomatosis were thought sufficient to illustrate this lesion Two more have been found in the office records.

The Author wishes to acknowledge the help of Dr Paul Ross, who has worked with him on many of the cases, also for his kind ness and that of Drs Gerald Peterson and Harold Gordon in proofreading the manuscript Dr Sam Black has rendered inval uable aid in the classification and histologic study of these cases Our colleagues, the roentgenologists, Drs Bell, Douglas, Peterson and Blackburn at the office, and Drs Johnson and Maxwell at the hospital, were ever ready and willing to discuss and work out the details of their fields, far beyond the usual call of duty The aid given by our anesthesiologist, Dr J A Bishop and the ever dependable nursing service both in the operating room and on the floor, has been greatly appreciated Miss Mary Weber my secretary worked patiently and faithfully in preparing the manu script and to her I am indeed grateful Mr Lawrence Jelsma was especially helpful in correcting the proofs. I am thankful to my wife for her encouragement and understanding of the time and effort required to gather together and write this work Mr Jack Hand prepared most of the illustrations

Last, but not least, the Author wishes to express his sincere appreciation and thanks to the Publishers for their ever helpful suggestions and guidance in the preparation of this work.

F J

Louisville Ky

CONTENTS

PRIMARY TUMORS OF THE CALVARIA

Chapter I

GENERAL CONSIDERATION OF CALVARIAL TUMORS

INCIDENCE

PRIMARY tumors of the calvaria are rather uncommon Percentagewise tumors seem to occur less frequently in bone which is ossified in membrane or primitive connective tissue, such as the bones of the calvaria

Case reports being scarce, the reports of a group or series of cases are more difficult to find In one such group of cases, Vandenberg and Coley[1] found that benign tumors of the calvaria comprised only 2.4% of a total of 1 580 bone tumors occurring in the body Geschickter[2] found this percentage to be even less in another series of 500 cases

It is quite likely however that these tumors occur more frequently than the literature would indicate, for various reasons Many small osteomas need no care. They are observed indefinitely, and are neither operated nor reported This happens to many small tumors that grow slowly and cause few symptoms

Searching through our records, we have found 44 cases of benign and 5 malignant primary tumors of the calvaria In most instances they have come to us because of a pain or because of an enlargement somewhere over the head which is often referred to by the patient as a knot The incidence would be higher than in a bone clinic where the lesion is found coincidentally when the patient is being studied generally or because of a bone tumor elsewhere The incidence is higher in the office where the patient is referred because of a particular symptom referable to the head than would occur as normal percentage per thousand of average populace

CLASSIFICATION OF TUMORS OF THE CALVARIA

In outlining a classification of tumors of the calvaria for clinical purposes, we wish to include along with the primary lesions, those tumors produced by general disease processes, if the incidence of the skull manifestation is relatively high in the symptom complex of the disease. So this would include some lesions that are not neoplastic in nature.

GENERAL DISCUSSION OF CASE SUMMARIES

Statistically speaking, the total number of tumors under consideration is not large enough to permit one to draw definite conclusions in regard to incidence as to age and sex. However, there may be sufficient evidence to suggest that the benign lesions occur more frequently in the female than in the male. In our group of 43 benign tumors, 72% occurred in the female. In Vandenberg and Coley's[1] series of 15 cases, 66⅔% occurred in females. The average age was 38 years.

The reverse is true in malignant cases, there being 80% of our cases in males. In Vandenberg and Coley's[1] series of 7 cases, 71% occurred in males. The malignant tumors, as one would expect, occurred in the younger individuals.

It is interesting to note that in our series and in that of Vandenberg and Coley[1] the vast majority of tumors occurred in the frontal bone. The parietal ranked second and the occipital a poor third. Only 2 tumors affected the temporal bone. Tumors of the base of the skull have not been included in the study.

The benign tumors, as a whole, were usually small. Most of them had been discovered some months or years before consulting a physician. The tendency for the lesion to grow slowly larger and in some cases to cause pain, was the common reason for seeking help. The usual complaint was that they had a knot or bump, which was painful or sore and it was slowly getting larger. Headache was only occasionally mentioned, and this especially if the dura was involved or the diploe expanded.

The malignant tumors were larger, more painful and grew more rapidly. The pain was quite severe in some cases. Headache was more common and intracranial signs and symptoms were present, depending upon the location and size of the lesion.

CLASSIFICATION OF TUMORS*

I BENIGN TUMORS	II. GENERAL DISEASE PROCESSES WITH TUMOR LIKE LESIONS	III. MALIGNANT TUMORS
Osteoma	Eosinophilic Granuloma	Osteogenic Sarcoma
Hemangioma	Xanthomatosis	Chondrosarcoma
Cavernous	Fibrous Dysplasia	Fibrosarcoma
Capillary	Paget's Disease	Ewing Tumor
Giant Cell Tumors	Leontiasis Ossea	Myeloma (plasma cell)
Epidermoid Cysts	Hyperostosis Frontalis Interna	Reticulum Cell Sarcoma
Traumatic Cysts		Hemangio-endothelioma
Chondroma		Chloroma
Lipoma		
Ossifying Fibroma		

*Each tumor is discussed in the order listed above

SYNOPSIS OF CASES

N	Name	Age	Sex	Location	Size	Symptoms	Path Diag	X-ray Diag	Surgery	Remarks
1	G M J 57	28	F	R.occ	3 × 3 cm	Lump	0	Osteoma	No	
2	C A L 57	54	M	L.fr	1 × 2 cm.	Knot	0	Ost.	No	
3	H D.S 51	22	F	R fr	2 × 2 cm	Knot 3 yrs. Headache	0	Ost.	No	
4	A A R 55	25	F	R.occ.	2 × 3 cm.	Pain 2 yrs	0	Ost.	Yes	Inner table left intact
5	B L B. 55	22	F	R.fr	2 × 1 cm.	Knot 3 yrs	Osteoma	Ost.	Yes	Inner table left intact
6	J B 58	36	F	R.fr	6 × 6 cm	Knot 6 yrs.	Ost	Ost.	Yes	Tumor removed-bone flap
7	J J 43	17	M	L.fr	6 × 7 cm.	Knot 10 yrs.	Ost (2)	Ost.	2 removed	Inner table left intact
8	C E C. 57	45	F	L fr	1 × 1 cm.	Knot 18 mos.	Ost.	Ost.	Yes	Inner table left intact
9	T.B. 58	36	F	L.fr	2 × 2 cm.	Knot 10 yrs.	Ost.	Ost.	Yes	Inner table left intact
10	P B 58	57	F	R fr	1.5 × 1.5 cm	No symptoms	0	Ost.	No	
11	N E.H 51	42	M	R par	1 5 cm × 2 cm.	Headache Pain	Cav Hemang	Hemang	Removal tantalum pl	1957—no removal
12	J L.S 52	51	M	R.fr	2 × 2 cm.	Headache	Cav Hemang	Hemang	Yes	Roentgenotherapy
13	M W 56	44	F	L.fr	3 × 2 cm.	Soreness	Cav Hemang	Hemang	Yes	OR and plastic cranioplasty
14	H C. 52	28	F	L. par	3 × 2 cm.	Headache	Cav Hemang	Eosin Gran.	OR and Roentgenotherapy	Tantalum cranioplasty
15	J M McM. 50	40	F	L par	2 × 2½ cm.	Headache 2 yrs	Cav Hemang	Hemang	OR and tant. cranioplasty	
16	R B. 54	71	M	L.fr	2 × 2 cm.	Headache	Hemang	Hemang	Yes	
17	R.T 54	35	F	L.fr	8 × 5 cm.	Headache		Cap. Hemang	No	Roentgenotherapy
18	A.H 55	66	F	R.occ.	2 × 3 cm	Throbbing pain	Cap. Hemang	Cap. Hemang	Ligated Ext. car	
19	B.S. 58	19	F	Occ.	7 × 5 cm.	Throbbing pain	A.V Fistula	0	Yes	Larger vascular mass removed Skull involved.

No.	Name	Age	Sex	Location	Size	Symptom	Path Diag	X-ray Diag	Surgery	Remarks
20	C F B. '58	48	F	L.par	4 × 5 cm	Headache	Epid cyst	Epid cyst	Yes	Injury 20 yrs before
21	C B. 44	27	M	R.fr	× 3½ cm.	Soreness	Epid. cyst	Epid cyst	Yes	
22	F W K. 47	23	M	R par	3 × 4 cm	Sore mass	Traum cyst	Epid		OR and plastic cranioplasty
23	G Y H 54	58	F			Headaches	Cyst (traum)		Yes	OR and Plastic cranioplasty
24	L.P 53	71	F	L par	3 × 3 cm.	Headache	Traum cyst	Epid	OR 55	OR and plastic cranioplasty
25	M L.F 58	4 mos	F	R par	3 × 4 cm	Tumor	Traum. cyst	?	OR 58	Cyst wall and outer table removed very vascular
26	K L. 50	71	F	R.occ	6 × 8 cm	Growing mass	Chond	Chond	Removal	
27	R M W 52	21	M	R.occ	1 6 × 2 cm	Headache	None	Eosin Gran	No	
28	C N C. 52	29	F	R par	1.5 × 2 cm	Soreness	Eosin. Gran	Eosin Cran.	Removal and tant. plate	Outer table eroded and soft tissue invaded
29	J Y 55	24	F	R par	2 × 2 cm.	Soreness	Eosin Gran.	Eosin. Cran.	Removal and plastic plate	Periosteum attached 9 eosin —WBC 17 000
30	E.S 54	35	M	R fr	1 × 2 cm	Pain	Eosin Gran.	Eosin Cran	Removal and plastic cranioplasty	Extended through pericranium
31	J.S B. 54	41	F	L.par	2 × 2 cm.	Local pain	Eosin Gran	Eosin Gran	Removal	Extended through both tables
32	W O 53	21	M	L.par	1 × 2 cm	Soreness	Eosin. Gran	Eosin Gran	No	
33	W R O 37	3	M	R.par	6 × 4 cm.	Pain	Xanthomatosis (2)	Xanthomatosis	No	Roentgenotherapy—multiple lesions
34	W L. 37	2	M	Fr & par	4 × 3 cm	Pain	Xanthomatosis	Xanthomatosis	No	Roentgenotherapy—multiple lesions
35	J R 58	20	M	Fr	4 × 5 cm	Pain	Fibr dyspl	Fibr dyspl	Yes	Plastic cranioplasty

No	Name	Age	Sex	Location	Size	Symptoms	Path. Diag	X-ray Diag	Surgery	Remarks
36	B.H 54	13	F	L fr.-par R fr par	5 × 5 cm. 2 × 3 cm.	Lump sore	Fibr dyspl (2)	Ost.	Yes	Cranioplasty Inner and outer tables expanded
3	P.J.H 50	17	F	R fr	4 × 5 cm. 3 × 3 cm	Headache dizzy	Mono-Fibr dyspl	Ost.	Yes	Cranioplasty Inner and outer tables expanded
38	C.H 45	61	M	Fr.-temp especially		Head enlarging		Paget's dis.	No	Circumference increased 8 in. in 10 yrs.
39	R H 45	55	F	Calvaria & base	Massive	Tumor 20 yrs.	Leon Oss.	Leon Oss.	Yes	
40	S A.S 57	16	F	L. zyg.-malar temp.	Large	L. face large		Leon. Oss.	No	
41	J B.H 39	55	F	Bi-fr		Headache		Hyperostosis fr Int	No	
42	E.S. 51	57	F	Bi-fr		Headache		Hyperos. fr Int.	No	
43	S.K. 55	43	F	Bi-fr		Headache		Hyperos. fr int.	No	
44	S R.M. 50	44	F	Bi-fr		Headache		Hyperos. fr int	No	Sella enlarged
45	R.S. 57	14	M	R.fr	3 × 2	Pain tumor site Headache	Osteog Sarc. (2)	Osteog Sarc. (2)	Extensive removal	Base involved also
46	C.P 48	14	M	R.occ. par	Grape fruit	Tumor mass	Osteog Sarc.	Osteog Sarc.	2 times in 5 mos.	2 ORS before we saw him
47	J G T 54	26	M	Bi-par	Lesion skull & other bones	1953 1956	Chondro-Sarc.	Chondro.	5 times in 3 yrs.	Multicentric chondrosarc. Surg and irradiation
48	C.H.B. 50	75	F	Bi-par	¼ × 1 cm	Headache	Pl. C. myeloma	Pl C. myeloma		Bone marrow positive B J pr
49	R.P 48	13	M	R.occ. par	Large	Headache	hemangio endothelioma	Mal.	Yes	Destruction of skull.

Only the unusual appearing osteomas were sectioned for microscopic study. The soft tissue of all the tumors otherwise was studied. In over 75% of the cases a tissue study was obtained. Forty four operative procedures were performed, there being multiple operations in 5 cases.

The exact diagnosis cannot always be determined before surgery by the clinical findings and roentgenographic interpretation. Sometimes a most exhaustive and deliberate study ends up with 2 or 3 possibilities. Thus the surgeon must depend upon his own experience, and during surgery utilize the help of a competent pathologist before deciding the type and extent of surgery to be employed. He may stop at times with only a biopsy and resort to some form of radiotherapy or other nonoperative treatment. A simple surgical removal may save extensive radiotherapy.

In some cases then surgery may not be indicated while in others the extent of surgery may be in question. Experience, surgical judgment and analysis of results of cases treated by various procedures as noted in scattered cases through the literature, all may point the way. But there are many problems of therapy that have not been resolved to date. These problems will be discussed with each particular tumor.

The problem of cranioplasty is usually decided by the type of tumor, the location and the extent of the cranial defect. In the benign cases, at the time of the initial surgery we have fitted a cranioplasty when the defect is large enough to require dural or cerebral protection or when cosmetic factors are a matter of consideration. It is easier to get a good fit using the one stage method of acrylic cranioplastic repair. In this way the exact thickness, shape and size can be obtained by molding the material before it hardens into the cranial defect.

CRANIOPLASTY

Plastic material has been used for many purposes during the past few years. Orthopedic surgeons[3] have used it for reconstruction of hip joints. It has been used by dentists. It has now become very useful in repairing skull defects. Rietz[4] reports that Zander in 1940 was first to use methyl methylacrylate for the repair of a cranial defect. Many neurosurgeons[5,6,7,8] since have reported

on its use for cranioplasty, and also concerning its favorable tissue reaction [9]

Plastic material is strong it is light and it is kind to the tissues The speed with which it may be prepared, and the ease with which it may be fitted into any shape of cranial defect, has made it especially valuable for this purpose

To Oliver and Blaine[10] and Woringer[11] should be given the credit of developing the one stage method of acrylic plastic cranioplasties This technique has made it possible at a moment's notice to mold a snug fitting plastic plate to fill any size defect Therefore, it can be used for repairing skull defects produced at surgery because of any reason or because of previous trauma, such as open depressed fractures or closed depressed fractures of the skull Rietz[4] has routinely used it in traumatic cases at the time of the initial surgery

We have used it since 1954 mainly for elective cranioplasties and for the repair of operative defects produced particularly in the removal of primary benign tumors of the calvaria We have had no complications to date, and we have used it in 21 cases We have obtained a more uniform fit and consequently there is no bulge or unevenness at the margin of the plastic plate, like sometimes one would have with even the best fitting tantalum plate This gives a better cosmetic result Furthermore, pneumograms will show normally and will not be obliterated as would occur with a tantalum plate in place Roentgenotherapy for any purpose can be used without interference by the plastic plate

The method we have used has been very satisfactory During surgery when the decision has been made that a cranioplasty should be used, the sterile resinous powder can be placed in a small mixing bowl and the sterile cranioplastic ampule of liquid can be added The nurse vigorously mixes the two substances, much as in making biscuit dough This mixing is continued for 3 or 4 minutes At this time the powder is thoroughly mixed and a doughy mass results. It is then taken up in the hands and worked pouring some Ringer s solution over the gloved hands to prevent sticking In about 6 or 7 minutes it will begin to feel warm and takes on a workable doughy form This is then molded into the defect over a piece of cottonoid that has been introduced to pro-



Fig 1a Showing outer surface of plastic plate molded to fit skull defect.
Fig 1b Showing under surface and flanged margin of plate Its contour varies with that of the skull to give a perfect fit

tect the dura and a thin flange is allowed to extend out over the periphery of the defect This will help stabilize the flap and safe guard against outside pressure forcing the plate inward Soon it will become warmer and before it has become hot, the plate is removed from the defect and allowed to cool pouring Ringer s solution over it Gradually it becomes fixed and at this time the finishing touches to secure a good fit are made When it has hardened the margins are trimmed with a rongeur If desired perforations for wire sutures are made at the margins of the flap

This plastic material looks like bone, it is about the same weight as bone, it feels like bone and it bites with a rongeur like bone

The cottonoid is now removed from the dura and the plate replaced A tight scalp closure is made and a mild pressure bandage is applied over the wound

A tendency toward unfavorable tissue reaction in animals has been reported from some laboratories So far as the writer has been able to determine no such reaction has been reported in practice and no doubt, many thousands of cranioplastic repairs

have been used throughout the country It seems likely that the possibility of untoward reactions is very minimal

In the malignant cases, a cranioplasty is usually not used Removal of the tumor will relieve the pain and headache, even though a cure cannot be effected Sometimes relief of suffering is sufficient to justify the surgery involved Certainly a cranioplasty is not indicated unless the end results will justify its usage.

BIBLIOGRAPHY

1 Vandenberg H J Jr Coley B L. Primary tumors of the cranial bones. *Surg Gynec & Obst* 90 602-612, May 1950

2. Geschickter C. F Primary tumors of cranial bones. *Cancer* 26 155-180 1936

3 Harmon, P H. Methylacrylate resins in surgery with special reference to the reconstruction of arthritic joints. *Mod Plastics* 20 56-57 pp 114-116 1942

4 Rietz, Karl A. The one stage method of cranioplasty with acrylic plastic. *J Neurosurg* pp 176-182 Mar 1958

5 Gurdjian E S Webster J E Brown J C. Impression technique for reconstruction of large skull defects *Surgery* 14 876-881 1943

6 Kerr A S The use of acrylic resin plates for repair of skull defects. *J Neurol & Psychiat* 6 158 1943

7 Small, J M Graham M P Acrylic resin for the closure of skull defects, preliminary report. *Brit J Surg* 33 106-113 1943

8 Woolf J I Walker A E Cranioplasty collective review *Internat Abstr Surg* 81 1 23 1945

9 Kleinschmidt, O Plexiglas zur Deckung von Schadellucken. *Chirug* 13 273-277 1941

10 Oliver L. C Blaine, G A new one stage method of cranioplasty with acrylic plastic *Med Pr* 220 167 169 1943

11 Woringer E. Schweig B Brogly G Schneider J Nouvelle technique ultra rapide pour la réfection de bréches osseuses craniennes à la résine acrylique. Advantages de la résine acrylique sur le tantale, *Rev neurol* 85 527 535 1951

Chapter II

BENIGN TUMORS OF THE CALVARIA

We have chosen to include in this group osteomas hemangiomas, giant cell tumors epidermoid cysts, traumatic cysts, chondromas lipomas and ossifying fibromas.

OSTEOMA

We have placed osteomas first because they are the most common of all the tumors of the calvaria The exact incidence is difficult to determine Many osteomas go unnoticed, also many are not brought to a physician s attention when noticed It would seem that our figure of approximately 20% would be too low

They have a tendency to occur in the frontal region[1, 2] and decidedly more frequently in females They do occur in all parts of the skull however including the basal area

There are many kinds of bony growths[3] but the lesions thought to be neoplastic in character are usually considered as being spontaneous, or possibly in some cases traumatic in origin In case of the calvaria, osteomas arise from the membranous bone and are very compact and eburnated They may be single or multiple large or small, but slowly become progressively larger It is thought that trauma may cause them to grow faster A rapidly enlarging tumor should be promptly removed

The patient is most often a young female but osteomas may occur in males and older people. They usually complain of a knot or bump on the head that has slowly enlarged The lesion may be smooth and round or irregular and flat The skin is mobile over the mass and there is usually no reaction of the scalp and no induration or discoloration Very few complain of pain tenderness or headache It would seem that if they did not increase in size, very few patients would bother about this tumor

The roentgenograms show a well circumscribed dense lesion, that extends outward Only infrequently is the inner table de

pressed or the tumor protruding inwardly. Often the inner table may be left intact at the time of removal of the tumor. There is no separation of the tables. The diploic markings fade away, as does the identity of the tables in the area involved by the tumor. Some large tumors present a spongy appearance. Increased vascular markings are not observed. There is an outward proliferation of bone and no absorption or erosion of bone. This is an important point of differential diagnosis from a meningioma.[4] Meningiomas also produce proliferation of bone, but the proliferation is mostly inward. Vascular markings are prominent and pressure signs are present. Spicule formation is common in meningiomas, but rarely found in osteomas. Hyperostosis frontalis interna is not difficult to distinguish from osteoma, because these lesions are less circumscribed, usually bilateral and extend intracranially. They usually occur in older patients.

Unless the osteoma has already grown to a large size, or is growing rapidly, one need not be in a hurry in deciding upon removal. The fact that they do steadily increase in size makes their removal desirable, however, this may be done at the patient's convenience. Their frontal location makes them readily visible on many people. The protruding nodule or mass may be quite troublesome to some patients. They may extend into the frontal sinus and at times block the outlet or cause other sinus problems. Thus we recommend that osteomas be removed.

The method of removal depends upon the type of lesion and its location. The usual small, circumscribed tumor can be removed by rongeurs and burr, leaving the inner table intact. A cranioplastic repair usually is not necessary. However, if it were advisable to remove the inner table with the tumor, a tantalum button or, if larger, a plastic plate can be molded into the defect.

If the tumor is large, a bone flap is turned about it. The dura and subdural space is inspected carefully for tumor or scar, and a plastic repair is made. However, we have found that in some cases the excessive tumor growth may be removed from the flap and the bone flap boiled for 20 minutes and then replaced. In these cases it is better to reflect the pericranium along with the scalp flap, so that nothing but the bone is boiled and replaced. We have found that this procedure gives a very good end result.



Fig 2. Case 1 R occipital osteoma.

There is no reaction and a good fitting bone flap replacement is obtained

One case, No 6 presented a rather unusual problem because of the size of the tumor (Please see later reference)

CASE REPORTS

In the report of all the cases to follow only the pertinent positive findings in the history physical neurological and laboratory examinations are mentioned This is done to save needless repetition of negative tests and examinations

Case 1 G M J female, age 31 seen in 1957 because of a lump 1 in. in diameter in the right occipital area. It was first noticed in 1949 and had been growing slowly since There had been no pain, but headaches of undetermined origin were present. Roentgenograms showed an area of increased density with bone proliferation extending 4 mm. above the surface of the skull. The diploe was less distinct and there was a tendency for the 2 tables to lose their identity Neurologic examination was entirely normal, so observation was recommended at the time So far removal has not been necessary



Fig 3 Case 4 R. occipital osteoma.

Case 2. C. A L. male, age 54 was seen in 1957 with complaints of fatigue, personality change, tenseness, disturbance of memory and concentration. He had no headache, but roentgenograms of the head disclosed a small osteoma in the left frontal region. This was not producing any symptoms and did not enter into the condition causing this man s troubles. It is being kept under observation.

Case 3 H D S female, age 22 was seen in 1951 because of a knot 1/2 cm. in diameter in the right frontal area This had been noticed about a year earlier was not painful or sore, but had grown somewhat larger Headache had been occurring for the past 7 months. There was a history of a car accident 3 years before and of being unconscious for 3 hours. Roentgenograms revealed a dense bone lesion thought to be osteoma Removal was suggested but patient declined She has not been seen since 1951

Case 4 A. A R. female, age 25 was seen in 1955 because of a knot in the right occipital area It had been noticed first 15 years before, and was slowly getting larger Only recently had it caused pain and was tender on palpation. The scalp was freely movable over it Roentgenograms showed a dense lesion extend

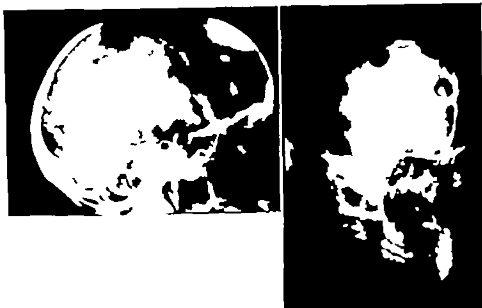


Fig 4a. Case 6 R. frontal osteoma.
Fig 4b. Case 6 Same case, after flap turned and tumor removed.

ing outward for about 1 cm. A little over a year later the tumor was removed, using a burr and rongeurs. The inner table was left intact. A small nerve passed directly over the tip of this lesion. It was sectioned There have been no further complaints from the patient, and she has been observed at yearly intervals.

Case 5 B. L. B female, age 22, was seen in 1955 because of a lump 2 x 1 cm. in the right frontal area. It had been noticed 3 years previously and had grown slowly It was not tender and caused no pain Tangential films showed a protrusion of 6 cm from the skull surface outward A burr was used to grind this tumor down to the inner plate The defect was saucerized a piece of gelfoam placed in it and the scalp closed in layers. There has been no evidence of further trouble from this lesion

Case 6 J B., female, age 36 was seen in 1948 because of a bump in the right frontal area It was noticed several years before and had caused no discomfort, but was gradually becoming larger At this time it measured 6 x 6 cm and protruded approximately 1 cm. from the surface of the skull. The inner table was slightly depressed A scalp flap was reflected within the hair line, including all of the pericranium, except that over

the tumor A bone flap was then turned about the tumor With chisel and rongeurs the protruding tumor was removed level with the surface. Then the flap was boiled for 20 minutes. During this time it was noted that the outer layer of the dura was slightly involved but the underlying brain was not affected. The bone flap was cooled and replaced and the scalp closed over it. Healing and convalescence were normal and subsequent observations since revealed the fact that the bone flap served as a good osteoplastic repair The bone flap fits perfectly is strong and boiling destroys the tumor cells. We have done this in cases where meningiomas involved the bone flap to a moderate degree and there has been no untoward reaction that we know of There have been fewer complications develop from the procedure than from the use of tantalum plates. The procedure may prove quite useful in certain cases

Case 7 J J, male, age 17 was referred to us in 1943 because of a knot in the left frontal area, attributed to an injury at 7 years of age, and another bump in the midfrontal area that resulted from a fall out of bed during infancy Both had been growing steadily Patient complained of headaches. There was no tenderness and the scalp was freely movable over both lesions. Roentgenograms showed the upper lesion to be denser than the lower one. It protruded 3 cm. from the surface of the skull, and measured 6 x 7 cm in diameter As osteomas frequently do the lower lesion involved the frontal sinus.

On 8/21/43 incision was made through eyebrow and the scalp reflected The osteoma was removed with burr and rongeurs, leaving the inner table, but including that part involving the outer wall of the frontal sinus The membrane of the sinus was not injured

On 9/16/43 the larger tumor in the midfrontal area was removed The outer portion was very hard Multiple perforator openings were made parallel to the skull surface, and the bone between rongeured away The inner portion was softer in places, but interspersed were very dense spicules that required a double action rongeur to dislodge With this method plus a curet, the entire tumor was removed The inner table of the skull was left intact.

We saw this patient again in 1952 He had served in the Army 6 years The skull contour was good and there was no evidence of recurrence of the tumor



Fig 5 Case 7 Osteoma, L. frontal and midcoronal margin

Case 8 C. E. C. female age 45 was first seen in April 1958 because of a nodule 1 x 2 cm in diameter in the lateral margin of the left supraorbital ridge It extended slightly into the orbital cavity It was not tender The patient had noticed this lesion for about 18 months and that it had increased rather rapidly in size Roentgenograms showed the area described above to be of increased density There was no absorption or erosion of bone and no increased vascularity This is a common site for osteomas. In May 1958 this lesion was removed with burr and rongeurs. The nodule protruding into orbital cavity was removed It consisted of dense eburnated bone. No repair of the skull was necessary and the contour of the supraorbital ridge was maintained Had this tumor been much larger and destroyed the supraorbital ridge a much more difficult problem of repair would have followed Thus, it is well to remove them early in this location, before they have attained a large size and involved the orbital ridge or the orbital cavity to any extent.

Case 9 T B C. age 36 female, was referred in 1958 because of a knot in the left frontal area It was 2 x 2 cm. in diameter



Fig 6a Case 10 Osteoma R. frontal area, lat. view
Fig 6b Case 10 Osteoma R frontal area, p.a. view

and protruded about 1 cm. out from the surrounding skull It had been present 10 years, becoming much larger in the past year or two When a child she was struck in this area with a brick. Roentgenograms showed expansion of the outer table of the left frontal bone almost to the midline. The inner table was not in volved No other abnormalities were seen

At surgery the periosteum had not been penetrated Bony proliferation extended in mushroom fashion out over the edge of the surrounding skull It was very hard With burr and curets the entire lesion was removed The inner table was not involved and left intact. The skull defect was saucerized

Case 10 P B age 57 female, was referred in 1958 because of constant headache, which however had been intermittent before the past 3 weeks. Complete physical and neurologic examination did not reveal any positive objective findings, except an area of increased density 1 5 cm x 1 5 cm in the right frontal area A slight elevation of the skull was palpated over this lesion It was not tender on pressure. The scalp was easily movable over it The headache apparently did not originate from the osteoma This lesion will be observed at intervals of 6 months or 1 year Should it cause symptoms or increase in size it should be removed

Unless otherwise stated the symptoms and complaints incident to the tumors in the above cases have been relieved The simple removal of the small osteoma has been adequate The larger tumors require the removal of a larger part of the calvaria and consequently the subsequent repair has been more extensive. The intracranial structures have seldom been involved

HEMANGIOMAS

These occur less frequently in bones than in soft tissue According to Wyke,[3] of 4 449 neoplasms of bone, as listed in 3 series of cases, there were only 36 or 0 7% hemangiomata After a thorough search of the literature, he found that hemangioma of the skull represented about 0 2% of bone neoplasms

There are 2 primary benign hemangiomas of the calvaria, the cavernous and the capillary types The capillary type occurs very infrequently It is more destructive, may be multiple and may produce symptoms of intracranial involvement as in the case of Haudousa [6] Holmes and Sweet[7] think that the cavernous type may result from the confluence of the capillary conglomerates In their 4 cases the tuber-frontalis was the area involved It is also the primary ossification center They thought this to be significant

In Toynbee s[8] original case, there was a tumor in each parietal area He also suggested that there may be some association be-



Fig 7a. Tangential view of hemangioma, showing radiation bone spicules gi
the typical sunburst appearance.

Fig 7b Tangential view of meningioma showing parallel spicules of
bony proliferation

tween the tumors and the centers of ossification of the parie
bone Considering the findings in the review of the cases availa
in the literature, there is little evidence to support this theory

Cavernous hemangiomas constitute the vast majority of the ca
reported This is the usual and classical lesion found in the calvar

Wyke[5] in 1949 reviewed the literature for the preceding 1
years and found only 39 cases of primary hemangiomas of t
skull In this group of 39 cases, most occurred in the fourth c
cade of life and most frequently in the parietal area For tho
who are interested all the authors reporting cases from 1845
1949 are listed in the above paper by Wyke[5]

Toynbee[8] has been given the credit for recording the first case of hemangioma of the calvaria Most of the subsequent reports have been of single cases. Bucy and Capp[9] found the skull involved in 18 cases Geschickter[10] reported 5 cases Vandenberg and Coley[11] reported 6 cases Rowbotham[12] reported 5 cases Holmes and Sweet[7] reported 4 cases in 1952

In our series there were 8 hemangiomas or about 16% of the total number of tumors One half occurred in the frontal and one half in the parietal area Two-thirds occurred in females

It was noticed that all complained of headache It is our thought that the tendency of the hemangioma to develop between the tables of the skull and ballooning of the tables may account for the complaint of headache In far advanced cases, the inner table may be eroded and the dura involved causing pain This is rare as they usually grow slowly The soft tissue of the scalp or dura is seldom involved So in palpation very little tenderness is encountered One can usually feel a slightly raised lesion involving the skull with the scalp freely movable over it In more advanced cases the outer table is penetrated and in this instance the pericranium is involved, producing local tenderness and soreness

Roentgenograms taken in the lateral anteroposterior and posteroanterior and tangential positions are very helpful in the study of the case Coley[13] mentions the sponge rubber or honeycombed appearance on the direct view The tangential films reveal in many cases a sun ray appearance Radiating trabeculae, like spokes of a wheel extend from a central point, while in meningiomas there is a tendency for the trabeculae to extend in a parallel fashion

There is neither shelving of the periphery of the tumor as in eosinophilic granulomas, nor the punched-out appearance However the radiolucent area is usually round or oval and is without increased vascularity of the surrounding bone. There is no hyperostosis Spreading of the 2 tables may occur with noticeable erosion This is greater in the outer than in the inner table In far advanced cases the outer table may be perforated but this is infrequent The inner table is less frequently penetrated than the outer table,[14] consequently the dura is not likely to be involved The above findings, involving the parietal bone in a female in

the fourth or fifth decade of life, would strongly suggest a cavernous hemangioma

These lesions gradually become larger, cause more headache, and some believe may even become malignant[14] We have no information or evidence which supports this idea However, it would seem that removal of the tumor surgically, offers a satisfactory method of handling these cases Roentgenotherapy alone or after surgery, has been used Surgery presents a definite diagnosis and removal under direct vision after the identity of the tumor is known One also escapes the effect of roentgenotherapy upon the deeper structures A cranioplastic repair can be done at the same setting, thereby obviating a second and later round of surgery

When exposed the tumor appears bluish in color with a hard shiny layer of bone forming the outer covering Beneath this outer layer the honeycombed bone is found Using a "sucker" one can completely empty the small honeycomb compartments and get a good view of its formation This part of the tumor is soft and can be removed with a curet or burr down to the inner part of the inner table. Sometimes the tumor has extended between the tables considerably farther than the roentgenograms would indicate The texture of the bone, as indicated by the rongeur bite, will help determine the lateral limits of the tumor Usually a burr rongeur and curet will be all that is needed for the removal of the hemangioma Bone wax will control the bleeding It can be placed on the burr while burning through the vascular part. Some choose to remove the lesion by bloc resection In this case a bone flap is turned about the periphery of the tumor If this is done, it is well to take into consideration that oftentimes tumor tissue extends far out between the 2 tables away from the raised and honeycombed central part

We have used roentgenotherapy in some cases, but more especially the capillary type In these lesions, therapy is repeated at intervals

The gross appearance is typical As mentioned before, the honeycombed area corresponds to the radiolucent area seen by direct roentgenographic examination When the pericranium and, if present the thin outer table is removed the lesion can be emptied



Fig 8a b External view of hemangioma showing honeycomb appearance (Courtesy B. D Wyke *Am J Roentgenol* and Charles C Thomas, Publisher)

of blood by suction Then the craters of various size can be easily visualized They seem to communicate, because suction in one place readily empties the other craters The blood appears to be venous It rapidly refills

Microscopically the cavernous type consists of lacunae lined with large well developed endothelial cells There may be connective tissue in places supporting the endothelial lining The bony trabeculae in some areas may be thickened while in other areas they may be replaced by fibrous tissue

Scheinberg and Elkin[16] recently called attention to a hemangioma of the skull associated with an angioma of the brain A case was reported showing the hemangioma of the occipital bone and an angiogram outlining the intracranial angioma No anatomical connection was demonstrated between the skull lesion and the intracranial angioma It isn t uncommon for hemangiomas to be multiple and to be located in different parts of the body

CASE REPORTS OF HEMANGIOMAS

Case 1 N E. H male age 42 was referred in 1951 because of headache for 2 years and frequent momentary dizziness during the preceding 2 months. There was a raised irregular area 1 5 x 2 cm in the right parietal area It was slightly tender Roentgenograms showed a discrete honeycombed lesion. There was no increase in vascular channels about it. An encephalogram did not reveal an associated intracranial lesion



Fig 9a (*Upper*) Hemangioma of occipital area.
Fig 9b (*Lower*) Same case with angiogram showing an angioma of the brain and no anatomical connection with the hemangioma of the occ. bone. (Courtesy L. Scheinberg, and M Elkin (the editor *Neurology* and publisher Lancet Publications)

The tumor was removed with a burr and rongeur It extended to the dura. A small tantalum plate was fitted Subsequent observations have revealed no further trouble

Case 2 J L S male, age 51 seen in 1951 because of headache and a knot in the right frontal area about 2 cm. in diameter Roentgenograms showed a circumscribed loss of density of the bone lateral to the frontal sinus and communicating with it. The inner table was not involved There was no increased vas-



Fig 10 Case 2. Hemangioma in R. frontal area.

cularity no hyperostosis. There was a faint honeycombed appearance in the direct view August 1952 the tumor was removed with burr rongeurs and curet, leaving the inner table. The frontal sinus wall was removed laterally but the membrane was not injured The microscopic sections showed the lesion to be a typical cavernous hemangioma

Postoperative roentgenotherapy was given because some tumor tissue may have been missed about the sinus. Subsequent films showed density of bone returning and the defect less prominent.

Case 3 M W female, age 44 referred in 1956 because of headache and soreness over a growth in the left frontal area for 1 year Three years previously she had been struck in this region Roentgenograms showed an area of decreased density involving the left orbital ridge and orbital plate There was evidence of bone destruction The direct film showed the honeycombed effect

In July 1956 the tumor was exposed It extended 1 cm above the outer table was very porous and filled with blood A sucker tip placed on one part of the tumor emptied the entire lesion of

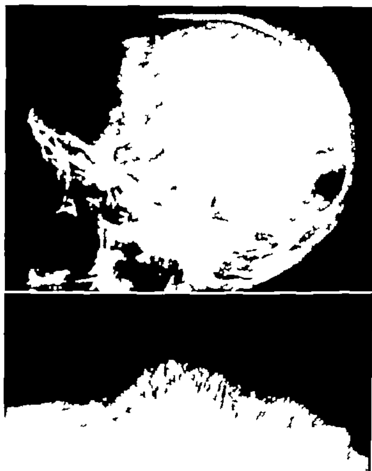


Fig 11a. Case 3 Lat. view showing radiolucent area above the left orbital ridge.
Fig 11b Same case, tangential view showing typical sunburst appearance of a hemangioma.

its blood content and one could see the sponge like construction of the tumor It was removed with rongeurs, along with involved bone surrounding the main tumor It was necessary to remove part of the ridge and roof of the orbit. After this a plastic plate was fashioned and secured with wire at 3 places. It gave a good cosmetic result The patient was seen as late as May 1958 and was free of symptoms

Case 4 H C. female, aged 28 was referred in 1952 because of headache and a lump in the left parietal area Roentgenograms showed an area of decreased density 3 x 2 cm. It was



Fig 12a. Case 4 Lat. view of hemangioma 1952.
Fig 12b Same pt. 5 years after operation—note another lesion developing posterior to first hemangioma



Fig 13 Case 5 Left parietal hemangioma.

elevated about 1 cm. above the skull surface and extended through both tables of the skull. There was no honeycombed appearance. The roentgenologist favored an eosinophilic granuloma because of the shelf like irregularities of the tables of the skull. However on April 2 1952 at surgery a typical honeycombed lesion was encountered The frozen section, and later histopathologic studies, proved this to be a hemangioma It was removed with burr rongeurs and curet Roentgenotherapy was given and later a tantalum plate fitted over the skull defect.

In 1956 headaches returned Just posterior to the tantalum plate patient complained of tenderness, and roentgenograms showed an area of decreased density One year later in November 1957 it was noted that this area had increased in prominence. There was a margin of normal bone between this lesion and the former defect, now covered by a tantalum plate. Roentgenotherapy was given The tantalum plate area was shielded. In the original films, a suspected lesion was noted in the site of the second tumor

Case 5 J M Mc female, age 40 was referred in 1950 because of headache and a lump in the left parietal area that was

Fig 14a Case 6 Lat view of frontal hemangioma
Fig 14b. Case 6 P.A. view of frontal hemangioma

first noticed 2 years previously The headache was quite severe. The lesion was 2 x 2 1/2 cm. in diameter and elevated almost 1 cm above the skull surface. The roentgenograms showed an area of decreased density but did not reveal the typical honeycombed effect found at surgery The density of the intervening bone forming the compartments must have been insufficient to show up in the exposure used It was removed with burr rongeurs and curet down to the dura and as far lateralward as necessary to encounter good normal bone A tantalum plate was fashioned and placed over the defect This patient has been observed at 1 to 2 year intervals and there has been no recurrence of the tumor There have been no complaints referable to her head

Case 6 R. B male, age 71 was referred in 1954 because of headache that had gradually become more disturbing There was a slightly raised tender area in the left frontal region. The roentgenograms showed a small oval radiolucent area in the left frontal region The tendency toward honeycomb formation was visible in the posteroanterior view This patient was treated with surgery and a plastic cranioplasty following removal of the tumor It was a typical cavernous hemangioma He was relieved of the headache.

Case 7 R T female, age 35 was referred in 1951 to us because of headaches, and at times a feeling of faintness for the past 4 years. There was a slight discoloration in the region of the left temple and cheek. We were able to locate roentgenograms taken in 1940 and 1946 The 1946 report noted there was thinning of the left supraorbital ridge and considerable extension of the lesion, as compared with the 1940 films. At this time roentgenotherapy was given and at intervals until 1951 at which time we first saw her Roentgenograms at this time showed destruction of the lateral margin of the left frontal sinus and superior margin of the orbital ridge and orbital roof Enlarged hard venous channels could be palpated in the left frontotemporal area. More roentgenotherapy had been given In 1953 and 1954 roentgenograms showed continued improvement, and patient improved clinically Whether or not this tumor had undergone malignant changes cannot be decided Certainly it destroyed more bone than the ordinary cavernous hemangioma and the blood vessels of the local area of the scalp became larger



Fig 15 Case 7 Radiolucent area in left supraorbital area Increased vascular markings. Cap. hemangioma

and later thrombotic, and in places calcified Many features of this case resembled a capillary hemangioma A section for microscopic study may yet be secured It is interesting to note that a hemangioma was removed in the area of the umbilicus when patient was an infant

Case 8 A. H female, age 66 was referred in 1955 because of a throbbing sensation in the right occipital area It was noted there was a fullness above and anterior to the mastoid Headache was moderate. The general physical condition was not good there being heart, kidney and gallbladder disturbances.

The roentgenograms showed a discrete area of increased vascular markings just above and anterior to the mastoid There was no honeycombed effect, no bone proliferation, but definite erosion A thrill could be palpated at times and pressure obliterated it At surgery a branch of the postauricular artery was

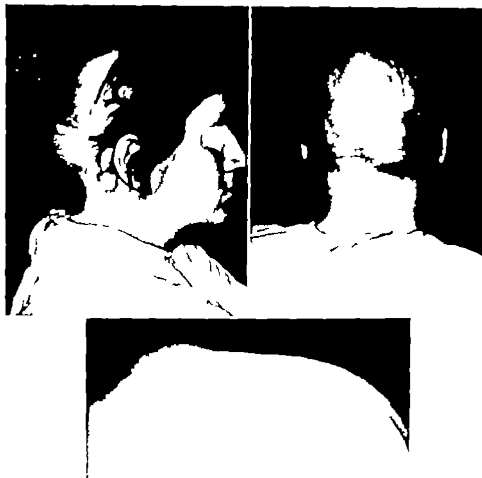


Fig 16a. (*Upper*) Case 9 Photograph of lat. view of arteriovenous aneurysm involving skull secondarily

Fig 16b (*Upper right*) Same case.

Fig 16c. (*Lower*) Tangential roentgenogram showing slight skull erosion. More extensive involvement found at surgery

found to be larger than normal and communicated with the lesion The vessel was ligated and sectioned and the local area in the skull removed with a burr rongeurs and curet. It was not necessary to remove the inner table of the skull. Neither was a cranioplasty required All symptoms were relieved The blood

vessels of this region were enlarged and tortuous No evidence of an arteriovenous fistula could be found

Case 9 This case is included because it is quite unusual and rare. It illustrates a point of differential diagnosis, viz a vascular tumor of the scalp producing secondary changes in the skull

B S., female age 19 was referred because of headache and a pulsating mass in the occipital area Five years before, she had struck her head on a bolt, while riding a school bus There were no ill effects immediately afterward except some tenderness locally A soft mass gradually appeared over the site of injury and a headache during the past year caused an increasing amount of trouble.

The mass was compressible, and as it was compressed the patient complained of pain By gradual compression the mass could be obliterated but when the pressure was released the mass quickly reappeared A pulsation and a high pitched sound synchronous with the heart beat were easily obtained but disappeared on compression of the right common carotid artery The size of the mass also became definitely smaller and again enlarged when the artery was released

Roentgenograms revealed some erosion of the skull Soft tissue films showed a mass containing some tortuous elongated densities thought to be distended vascular structures

At surgery greatly dilated vascular structures were encountered and removed en masse The outer table of the skull was eroded and in places the irregular surface of the skull was lined with thin endothelial lined structures, surrounded by considerable amount of fibrous tissue The pathologist reported the findings consistent with changes resulting from an arteriovenous aneurysm Clinical evidence would support this diagnosis.

GIANT CELL TUMORS OF THE CALVARIA

Benign giant cell tumors are commonly found involving the intracartilaginous bones of the body Membranous bones are seldom involved Pancoast, Pendergrass and Schaeffer[17] found 6 cases in the literature that developed in the bones of the calvaria Lichtenstein[18] in his book on Bone Tumors stated that he had never encountered a case originating in the calvaria Of 124 cases of giant cell tumors discussed by Coley[19] in 1950 none were in the calvaria Vandenberg and Coley[11] in 1950 reported a case

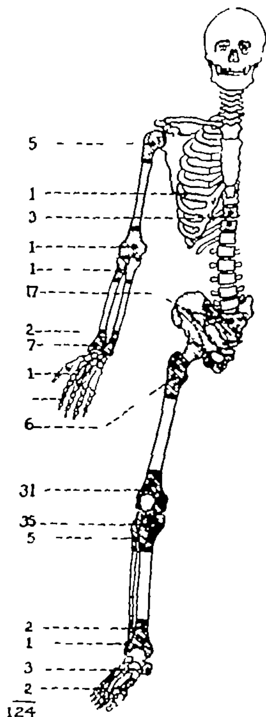


Fig 17 Showing location of 124 cases of giant cell tumors (from *Neoplasms of Bone* Courtesy Bradley L. Coley M D author and Paul B. Hoeber Inc.—publisher)

that mostly involved the basilar area, but the zygoma and portions of the temporal bone were invaded. This patient was treated with high voltage therapy, and 8½ years later there was no evidence of recurrence of the tumor. McNerney[19] recorded 2 cases, 1 arising in the temporal bone and 1 in the basilar area. Surgery and roentgenotherapy were both used with good results.

Trauma[20] and subsequent bleeding with reactive changes have been advanced as a possible causative mechanism. Bone rarefaction and destruction have been found roentgenographically. Diagnosis is made on histologic sections. The prognosis is good in spite of the tendency of the tumor to recur.

INTRADIPLOIC EPIDERMOID CYSTS

There are 2 kinds of epidermoid cysts which involve the head: the intradiploic and the intracranial type. The intradiploic type is a rare primary lesion of the skull beginning in the diploic region expanding[21] and eroding both tables as it develops. It may rupture through either or both of them and may attain a considerable size with relatively few symptoms. Except for the frontal and basal areas, the expanding tendency of the cyst produces a sclerotic appearance in the bone about the periphery of the lesion.

This lesion is not of infectious origin and not to be confused with the cholesteatoma found about the ear. It is thought to be congenital in origin. It grows slowly but may become quite large. It is soft and yellow in appearance and is composed of tissue derived from ectoderm. According to Ritvo[15] there are some supporting tissue, fatty material, cholesterol crystals and masses called "wood cells."

Netsy, Tolgia and Alexander[22] recently presented a Unitary Concept of Epidermoid Tumors of the Nervous System. They consider all of these tumors to represent embryological rests. They were also able to produce a typical epidermoid cyst by closing a portion of scalp beneath the suture line of the skin.

Treatment is surgical removal and cranioplasty.

CASE REPORTS

Case 1. C. B., male, age 27, was first seen in 1944 because of a raised place in the right frontoparietal area. It had been

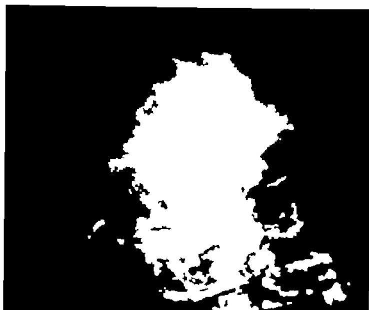


Fig 18 Case 1 Showing partial eburnation of bone about margins of an intradiploic epidermoid cyst of the skull

growing slowly over a period of several years. There had been some pain and headache during the past year The lesion extended about 1 cm above the surface of the skull and was about 2 x 3 1/2 cm. in diameter It was firm but not hard The margin of the skull about it could be palpated at one point. Roentgenograms clearly showed this skull defect, which seemed to involve both tables. There was slight eburnation of bone about the margin of the lesion The cyst was confined beneath the pericranium but had destroyed both tables of the skull. It was much larger than the bony defect would indicate Total removal was effected by sharp dissection and curet, using rongeurs to remove spicules about the margins of the defect. The pathologic report was, epidermoid cyst.

Case 2 C. F B female, age 48 was referred in 1958 because of rightsided headaches that had started in March 1958 At times they had been accompanied by nausea, watering of the eye and some blurring of vision In 1928 the patient had injured her head in the left parietal region. The scalp was lacerated at the time but she was not rendered unconscious.

The general physical examination and neurological examination revealed no abnormal objective findings, except in the

Fig 19a (*Upper*) Case 2. Epidermoid cyst of calvaria, lat view
Fig 19b (*Lower*) Case 2. P.a. view

left parietal region, where one could feel beneath the scar an irregular defect in the skull Roentgenograms showed a large irregular radiolucent area 4 x 5 cm. in diameter There was definite eburnation of the bony margins There was a slight expansion of both tables about the periphery and a destruction of both tables in the center of the lesion.

It was noted at surgery that the cystic tumor had destroyed the external table and was only covered by the pericranium at its central area It consisted of a soft yellow mushy material, typical of epidermoid cyst. The internal table had been eroded completely The tumor was adhered to the dura which remained intact. The outer and inner tables were expanded and thin The margins of the skull presented a concave U like" edge, the inner surface of which was so compressed that it presented a hard glistening eburnated appearance. A curet made no impression upon it, except to remove a fine membranous lining

The shelves of bone were removed by rongeurs, as well as the eburnated bone. A plastic plate was fitted and the scalp closed in layers.

The headache on the right side may have been caused by the tumor but it was thought to be a histamine type cephalgia

It was thought also that the injury 20 years before may have resulted in an inclusion of a piece of skin beneath the scalp and subsequently the cyst developed On the other hand the lesion quite likely developed from an intradiploic embryonic rest and the injury was noncontributory

TRAUMATIC CYSTS OF THE CALVARIA

Cysts of the calvaria are most often traumatic in origin They may involve the outer table, the diploe or the inner table wherever blood may collect and undergo organization If the injury results in a collection of blood under the pericranium a cyst may develop externally but it would gradually affect the outer table and would show up as an area of decreased density with a soft swelling overlying it Should a clot of blood develop and organize within the diploe, there would be a gradual eroding and expanding process that would be difficult to differentiate from an intradiploic epidermoid cyst. This was the diagnosis favored by the roentgenologist in 3 of our cases Epidural collections of blood

Fig 20a Case 2. Epidermoid cyst, showing tumor tissue removed.
Fig 20b. Case 2 Epidermoid cyst showing skull defect and plastic plate.

associated with fracture of the skull or trauma otherwise, which go on to organization may produce changes in the inner table of the skull A leptomeningeal cyst may result from tears in the arachnoid and dura, with fluid becoming trapped and pocketed Presumably the pulsation of the brain[14] causes a bony change of a cystic appearance Loyal Davis[13] discussed the etiology and reported 3 cases in 1934

The general appearance roentgenologically is that of an area of decreased density with eburnated margins. Sometimes the original hematoma is partly calcified, giving a flecked appearance.

Evacuation of the cyst is the treatment of choice. Sometimes a cranioplasty is necessary

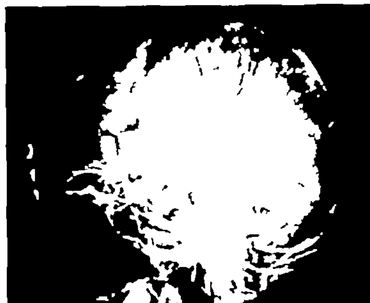
CASE REPORTS

Case 1 F W K. male, age 23 was referred in 1947 because of a headache, a pressure feeling dizziness and at times blurred vision. His symptoms were first noticed about 1 year previously In a car accident 7 years earlier he had injured his head The head was slightly larger than normal There was a tender raised place 3 x 4 cm in the left frontal area The scalp was freely movable over it. The roentgenograms showed a radiolucent area One roentgenologist suggested that the lesion lies between the tables of the skull and that ' the inner table was depressed The margins were all eburnated especially the inner table, suggesting to the roentgenologist an epidermoid cyst of the skull

At surgery the pericranium was not attached A bone flap was turned about the tumor The flap measured 4 cm. at its thickest point. The dense inner plate was removed to expose a cystic cavity containing some spongy bone and fibrous tissue. Both the dura and arachnoid were thickened Microscopic study showed some chronic fibroblastic proliferation and a few lymphocytes. A tantalum plate was fitted This patient has been observed on several occasions in the past 10 years and has not complained of further symptoms *

Case 2 G Y H female, age 58 was referred in 1954 because of a tender swollen place 3 x 4 cm. in diameter in the right parietal

*A recent very interesting case of intradiploic traumatic cyst has been added—see page 106



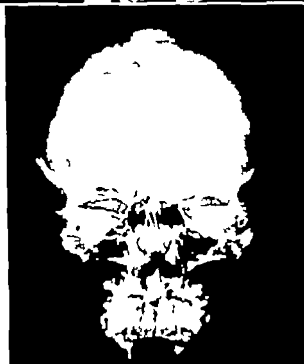


Fig 21a. Case 1 Lat view Intradiploic traumatic cysts of the left frontal area cortical markings diminished over fr lobe.
Fig 21b Same case, p a. view

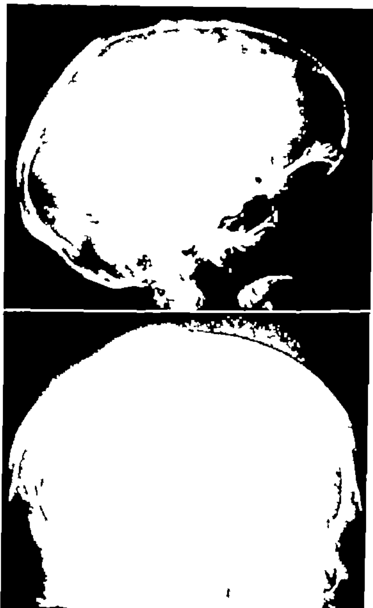


Fig 22a. Case 2. Traumatic leptomeningeal cyst, lat view
Fig 22b P A view same case

area She remembered striking her head in this area several times. At one time she was unconscious for several days. She had been aware of a slow growing swelling during the past 4 years The neurological and physical examinations did not reveal anything abnormal However the roentgenograms showed

an area of decreased density 3 x 4 cm in diameter raised 2 cm above the skull surface The churnated margins gave the appearance of an expanding process between the 2 tables of the skull Again, the radiologic diagnosis was that of an epidermoid cyst

At surgery when the pericranium was removed from the tumor the bony shell appeared a reddish brown The texture was porous and by actual count 20 drops per minute ("crocodile tears") of spinal fluid exuded through the tiny openings of the hard outer plate of bone The removal of this plate uncovered a cavity filled with cerebral spinal fluid and a small piece of pedunculated cerebral tissue which extended through the inner table and dura The bony floor of the cavity was removed about the protruding cerebral tissue and the dura opened The brain tissue was normal beneath and extending from the surface was this pedunculated piece of normal appearing brain about 1 cm. in its greatest diameter The pedicle was sectioned Microscopic sections showed it to represent highly vascular cerebral cortex. The dura was closed tightly and the cavity filled with gelfoam and a plastic plate molded and secured in place with wire sutures. The patient has been observed on several occasions since and has had no complaints The skull contour is good There has been some thought about this lesion being of congenital origin, rather than of traumatic origin

Case 3 L. P age 68 was referred in 1955 because of a constant headache for several years and a cranial defect in the left parietal area about the size of a half dollar She had in the past 3 years consulted many physicians and tried various medicines and procedures to obtain relief of the headache. Roentgenograms taken 1 and 2 years previously were compared with the the ones taken in 1955 There was definite enlargement of the area of decreased density The radiologist reported that the area of destruction, 3 x 3 cm in diameter lying in the left post parietal region 2 inches from the midline, appears to involve predominantly the diploic portion of the bone. There is more involvement of the inner table than the outer table. There is some sclerosis of bone about the periphery suggesting that it may have been present for an appreciable length of time He then suggested a diagnosis of an epidermoid cyst She had suffered a rather severe injury to the head in 1941

At surgery we found a leptomeningeal cyst. The thin outer shell of hard bone was removed exposing a cavity filled with cerebrospinal fluid The inner table was absent. There was a dural opening about 1 cm. in diameter extending into the subarachnoid space, and through which some large blood vessels protruded Cerebrospinal fluid ebbed and flowed with each pulsation The blood vessels were replaced beneath the arachnoid and the dura was repaired using a flap from the outer layer A piece of gelfoam was placed in the cavity and a plastic plate molded to fit the skull defect.

In the past 3 years she had done very well The old headache has gone and she has resumed her usual household functions.

Case 4 M F age 4 months, first seen August 5 1958 because of a lump on the back of the head It was noted immediately after birth, at which time it was soft. Since, it had gradually become smaller hard and fixed. It was 4 x 3 cm. in diameter and protruded about 1 cm above the surface of the skull in the right parietal area. It was not tender and the scalp was readily movable over it.

Roentgenograms showed a bony projection in the right parietal area, which seemed to have developed over the outer table of the calvaria It was 3 x 4 cm. in diameter and extended above the outer table 1 cm. The outer table was distinct and was not depressed There was an area of decreased density between the outer table and the dense outer layer of the tumor which was thicker than the skull and appeared almost as dense. The layer of decreased density between represented part of the hematoma that had not yet become calcified and may well in the future have become cystic and develop into a typical traumatic subpericranial cyst.

At surgery the pericranium was reflected from the tumor There was a blue discoloration of the entire tumor The hard glistening outer layer was completely removed with rongeurs. Each bite produced fresh brisk bleeding Bone wax was used after each bite to control bleeding This blue vascular bony outer layer was removed exposing a normal under layer of bone. A cranioplasty was not necessary The pathologist reported that the findings were indicative of a traumatic cyst of the skull

Fig 23a. Case 4 Lat view of R. parietal subpericranial cyst (traumatic)
Fig 23b Tangential view of cyst, same case.

CHONDROMA

I have been unable to find a report of a chondroma of the calvaria although I am certain that some must have been recorded In our records we have found 1 case involving the occipital bone, also a slowly growing intradural chondroma, extending from lumbar 1 to sacral 2 area It had completely entangled the cauda equina from the conus medullaris to the sacrum. Apparently from the history given its development had extended over a period of 25 years Copeland[1] states that chondromas are seldom multiple, and are usually found in the small bones of the hands and feet, the sternum spine and ribs, and occasionally in the long bones Coley's[13] diagram shows the distribution of 24 cases

They grow slowly but may become quite large in time There is little discomfort or pain On examination the lesion may seem much harder than when exposed at surgery There may be a tendency to gain the impression from palpation that the tumor might be an osteoma Also there is a tendency for the clinician to group these lesions with benign osteochondromas I have encountered the latter in the parasellar region of the head They tend to develop in the basilar areas

Chrondromas are benign but invasive, and definitely recur after incomplete removal Some may undergo malignant changes. Complete study of the specimen removed should be made before ruling out malignancy

Roentgenologically they produce a radiolucent effect, and many times a cystic appearance If sizable, there will be a soft tissue shadow outlining the tumor In the case reported below there was both an area of decreased density of the skull and a large soft tissue shadow Lichtenstein[19] stresses the importance of spotty calcification or a calcific stippling within the translucent areas

CASE REPORTS

K L female, 71 years of age, was referred to us in 1950 because of a large tumor in the right postauricular region. She reported that approximately 30 19 and 11 years previously a cartilaginous tumor had been removed from the same place It was noted in the last hospital record that the surgeon made

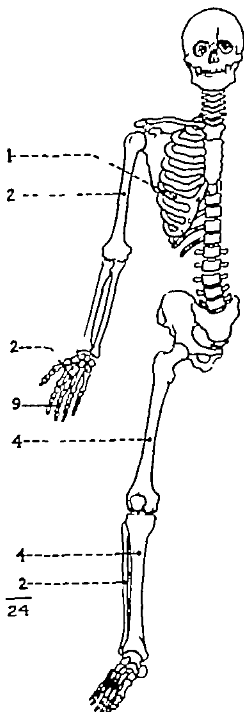


Fig 24 Showing distribution of 24 cases of central chondroma, (from *Neoplasms of Bones* Courtesy Bradley L. Coley M.D author and Paul B Hoeber Inc., publisher)

a presumptive diagnosis of an osteoma but his surgical note indicated the tumor was much softer than he had expected, and changed the postoperative diagnosis to a chrondroma

The mass was about the size of a hen's egg The patient was concerned mainly because of the size of the tumor It was not tender and was not movable. It was encapsulated with tissue moving freely over it The roentgenograms showed a hazy area of decreased density with some areas of spotty calcification. Tangential films showed a large tissue shadow representing the protruding tumor There was no increased vascularity

At surgery as had been formerly noted the lesion was firm *but not hard It was an encapsulated nodular pearly white* tumor about 6 x 7 cm in its greatest diameter It was removed with a sharp chisel after which a burr was used to remove the involved bone, which we thought to be the parietomastoid region. Part of the parietal bone and a portion of the posterior margin of the mastoid bone was involved It was not necessary to remove the inner table

The microscopic sections showed closely packed cells, with some rounded nuclei, definitely distinct cartilage cells. In some areas the cells were more scarce and the intercellular substance appeared collagenous. The pathologist kindly checked the slides of 1939 and found the appearance to be essentially the same, except the latter was a little more anaplastic. Both were benign. To date the lesion has not recurred

OTHER BENIGN TUMORS

In this category we have chosen to place primary lipoma and osteogenic fibroma These tumors occur very *infrequently in the* skull but should be mentioned nevertheless

LIPOMA

Lipomas may develop anywhere that one can find fatty tissue They may occur within the head along the brain stem the hypothalamus the corpus callosum and inside the ventricles We have removed them from within the dura in the lower lumbar area *They infrequently occur within the medullary canal of long bones* Smith and Feinberg[24] recently reported a case of intraosseous lipoma of the fibula and stated that only 1 other intraosseous lipoma had been reported in this country [25]

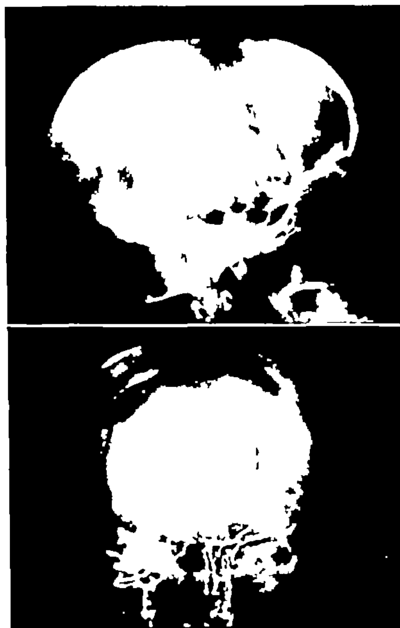


Fig 25a Lat view of a primary lipoma of the skull
Fig 25b P.A view (same case) (Courtesy Dr Loyal Davis and Lea & Febiger publisher) from case record of Dr Davis

As a primary tumor of the calvaria we know of but one case, that reported by Loyal Davis [24] Through the kindness of the above author who kindly sent the entire case record we have

been permitted to use this rare and remarkable case of primary lipoma of the calvaria

This patient female, age 42 was first seen by Dr Davis in 1935 A mass had first been noted in the left frontal area in 1915 In 1921 it was removed but gradually grew back, until in 1935 it measured 6 5 cm. in diameter and protruded 1 5 cm. from the skull surface. It was firm throughout, except the vertex, where there was a soft irregular area possibly the former operative defect. There was no evidence of increased intracranial pressure, or focal signs of cerebral dysfunction The roentgenograms showed a large area of increased density some bone proliferation and some bone destruction.

At operation it was found that the tumor had grown through the skull at the soft spot over the tumor vertex Burr holes were placed about the periphery of the tumor Here the skull was almost an inch thick and quite vascular The bone and tumor were removed The dura was intact The brain beneath was not involved.

The tumor was encapsulated yellow but not lobulated The operator mentioned in the operative note that it resembled fat, although he did not know at the time it was a fatty tumor

Microscopic studies showed this tumor to be a typical lipoma

OSSIFYING FIBROMA

Ossifying fibroma is mentioned by Copeland [1] We have never encountered one. He thinks they are quite rare, occur at times in the calvaria and are benign Excision is recommended

BIBLIOGRAPHY

1 Copeland M M Benign tumors of bone *Surg Gynec & Obst.*, *90* 697 712 June, 1950
2 Young B R Roentgen findings in tumors of the skull. *Radiology* *55* 321 326 Sept. 1950
3 Meredith, J M Osteomas of the cranial and base 3 cases illustrating 2 varieties. *South Surg* *16* 785–795 Aug 1950
4 Rand C. W Osteomas of the skull report of 2 cases 1 being associated with a large intracranial endothelioma *Arch Surg.*, *6* 573–587 1923
5 Wyke, B D Primary hemangioma of the skull a rare cranial tumor *Am J Roentg* p 302, March, 1943

6 Haudousa A S Multiple capillary hemangioma of the skull bones *Brit J Surg 31* 326 Jan 1947
7 Holmes, E M Sweet W H et al Hemangiomas of the frontal bone *Ann Otol Rhinol & Laryngol 61* 45 March, 1952
8 Toynbee J An account of 2 vascular tumors developed in substance of bone *Lancet 2* 676 1945
155-180 1936
9 Bucy P C Capp, C S Primary hemangioma of bone with special reference to roentgenologic diagnosis. *Am J Roentgenol 23* 1 33 1930
10 Geschickter C. F Primary tumors of cranial bones. *Cancer 26* 155-180 1936
11 Vandenberg H J., Jr Coley B L. Primary tumors of the cranial bones *Surg Gynec & Obst 90* 602 12, May 1950
12 Rowbotham, G F Hemangiomata arising in bones of the skull, *Brit J Surg 30* 1-8 1942.
13 Coley Bradley L. *Neoplasms of Bone* Hoeber New York, 1949 p 423
14 Abbott W D Angioma of the skull. *Ann Surg 106* 1100-1105 Dec. 1937
15 Ritvo Max *Roentgenographic Diagnosis of Diseases of the Skull* New York, Hoeber p 353
16 Scheinberg L Elkin, M Hemangioma of the skull associated with intracranial angiomata *Neurology* Aug 1958 p 651 (fig 2 and 3)
17 Pancoast, H K Pendergrass, E. P Schaeffer J P *The Head and Neck in Roentgen Diagnosis* Springfield Thomas, 1940 p 247
18 Lichtenstein L. Jaffe, H L Fibrous dysplasia of bone. *Arch Path 33* 777 1942.
19 McNerney J C. Giant cell tumors of bones of the skull *J Neurosurg 6* 169 1949
20 Copeland M M Geschickter C. G Tumors of the Bones and Joints. Chapt 28 pp 1121 1145
21 Alpers, B J Harrow R Cholesteatoma originating in the skull bones, causing symptoms of intracranial pressure. *Am J Surg 18* 51 57 October 1932
22 Netsky Tolgia and Alexander Personal communication.
23 Davis, Loyal Chorbosku, J Cyst formations of the skull *Surg Gynec & Obst 58* 12 31 January 1934

24 Smith, W E, Fienberg R Intraosseous lipoma of bone. *Cancer 10* 1151 1152, 1957
25 Dickson, A B Mason, M W Noller W R. Lipoma of bone of osseous origin *J Bone & Joint Surg 33A* 257 259 1951
26 Davis, Loyal *Neurological Surgery* Philadelphia, Lea & Febiger 1936 p 193 Also personal communication

Chapter III

TUMOR LIKE LESIONS OF THE CALVARIA
(DUE TO GENERAL DISEASE PROCESSES)

We have chosen to place in this category those tumors produced by general disease processes if the incidence of the skull manifestations is relatively high in the symptom complex of the disease. In this group we have included eosinophilic granulomas, xanthomatosis, fibrous dysplasia Paget's disease, leontiasis ossea and hyperostosis frontalis interna Also, one could include craniotabes of Ricketts, cranial changes in Gaucher's disease, osteoporosis circumscripta osteitus fibrosa cystica, and syphilitic skull involvement. The latter group does not represent as much a local cranial problem as a general systemic problem which is usually more compelling than the incidental cranial lesion Also the cranial manifestation of these diseases needs no particular local form of treatment

EOSINOPHILIC GRANULOMA

In 1940 Otani and Ehrlick[1] first applied the term eosinophilic granuloma to the lesion involving the skull It is characterized by a single or multiple punched-out areas of lesser density usually in females below 30 years of age The cause is unknown but several writers[2,3] associate this lesion more or less with xanthomatosis Clinically the 2 diseases present an entirely different symptom complex The symptoms, age, incidence and the skull pattern of the 2 respective lesions are entirely different except for an occasional case, such as reported by Swaine and Williams [4] The course of the 2 diseases is different as one can note from comparison of the cases listed below Knighton and Fox[5] thought it to be a primary disease of the reticuloendothelial system Shuster and Flynn[6] inclined toward the infectious origin

The patient is usually a female In our cases, 50% were females The majority of the patients are in early adult life. They complain of pain and soreness over a local area, most often the parietal region This is due to the fact that there is a tendency for this lesion to grow outwardly and involve the pericranium and even the muscles, or at times the dura[7] inwardly We have noticed an increase in the eosinophils in the blood count. In our cases, the patient has not been ill and has not presented systemic or general complaints as in cases of xanthomatosis

Roentgenologically, the lesion presents an irregular punched out area of decreased density, with no increased calcification or eburnation about the borders It may suggest a metastatic lesion, especially if there are multiple areas Other bones of the body may be involved There is no sunburst appearance on tangential views, as in the case of a hemangioma These lesions are usually smaller than hemangiomas, when first seen It may be that they cause pain earlier A soft tissue shadow may be seen if the lesion has extended beyond the outer table They are much different from the large multiple map-like configurations of Schuler-Christian s Disease The late Clyde McNeil[8] noted a double shadow which he thought characteristic of the disease. This he attributed to different degrees of involvement of the 2 tables of the skull

At surgery one finds a granulomatous lesion confined beneath the pericranium or perhaps growing through into the soft tissues. We have seen it invade the temporal muscle quite extensively Also it may involve the dura or extend through the dura and produce adhesions or direct cerebral involvement.[9]

The gross specimen may be cystic in places and may have hemorrhagic areas, but is soft and does not have the honeycombed sponge-rubber effect of the hemangioma It is less vascular *Microscopically the diagnosis is not difficult.* There are foamy appearing large mononuclear cells with eosinophils scattered or in clumps There may be some plasma cells and lymphocytes. Later connective tissue and new bone formation may be seen

We feel that a diagnosis cannot at times be made without a biopsy Also there is dural and intracranial involvement at times. Consequently surgical removal of these tumors is advisable. Cer

tainly removal and plastic repair will effect a cure Some advise doing nothing Some suggest radiotherapy [3]

The surgical plan we employ is to remove the lesion with a curet and sharp dissection, including any of the scalp that is in volved The dura is saved, if possible, but the tumor tissue is removed Additional bone is removed with rongeurs if necessary, and a plastic repair of the skull is fitted in place

In this study of cases, there were 6 patients with eosinophilic granuloma or about 12% of the total The lesion occurred most frequently in the parietal area There were no multiple lesions In 3 of the 6 cases the lesion had invaded the soft tissue of the scalp and the dura in 2 cases Pain was more prominent and more severe in these cases, probably because the tumor more frequently involved the scalp and dura Elevated eosinophil count was noted in most of the cases No other systemic changes were noted

CASE REPORTS

Case 1 R M W male, age 21 was referred in 1942 because of headache for 6 months and because of a knot 1 6 x 2 cm in the right occipital area The mass was tender on pressure. Roent genograms revealed an area of decreased density with shelf like margins. The outer table of the skull was destroyed We do not know what became of this patient.

Case 2. C. A C. female, age 29 was seen by us in 1952 be cause of pain in the right parietal area There was a very tender palpable nodule that the patient had noticed for 5 weeks. The blood Kahn was positive, but the spinal fluid was normal in all respects. Roentgenograms showed an area of destruction near the right lambdoidal suture involving both tables. It was 1 5 x 2 cm in diameter There was one small fleck of bone in the central portion of the defect that had not been completely absorbed The roentgenologist thought it was characteristic of an eosin ophilic granuloma There were 2 eosinophils and the sedimen- tation rate was 20 Ribs, heart and lungs were normal.

At surgery the tumor was encountered out in the galea. It was removed with sharp dissection and curet down to the dura A tantalum button was fitted in the defect The pathologic report was that of an eosinophilic granuloma A yearly follow up since has revealed no further complaints.

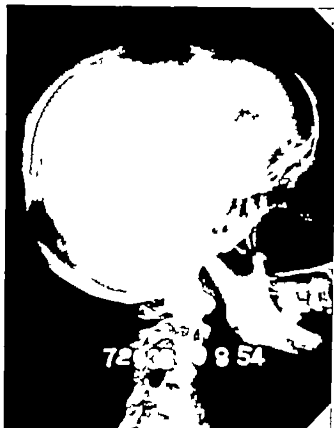


Fig 26a Case 4 Eosinophilic granuloma R. frontal.

Case 3 J Y female, age 24 was referred in 1951 because of pain for several months in the right side of the head gradually getting worse. Palpation revealed an extremely tender mass 2 x 2 cm. in the right parietal area The roentgenograms showed a sharply demarcated defect in both tables of the skull, thought to be an eosinophilic granuloma The eosinophil count was 9 and the sedimentation rate was 7

At surgery the tumor had extruded through the outer table. It was removed by sharp dissection, curet and rongeurs. The dura was involved but not penetrated A tantalum button was placed in the skull defect. The microscopic sections showed it to be eosinophilic granuloma Roentgenograms of the skull taken in 1958 did not reveal any new lesions and the patient had no complaints.



Fig 26b Same case—low power
Fig 26c. Same case—high power

Case 4 E S , male, age 35 was referred in 1954 because of sharp pains for 2 months in the right frontal area A tender nodule could be palpated The roentgenograms showed a de fect in the skull involving both tables and thought to be an eosin-ophilic granuloma The eosinophil count was 1

It was removed with curet and rongeurs down to the dura Part of the involved dura was included The external portion of the tumor had penetrated the pericranium and a large por tion was located in the muscle. A plastic button was placed in the defect of the skull Microscopic sections revealed a typical eosinophilic granuloma There have been no further com-plaints when last seen in 1957

Case 5 J S B female, age 41 was seen in 1955 because of severe pain in the left parietal region This had been present for 6 months but had gradually increased Roentgenograms re vealed an area of decreased density 2 x 2 cm in diameter It was less sharply defined than usual but considered as an eosin-ophilic granuloma

At surgery the tumor had involved the soft tissue of the scalp and dura It was completely removed and the defect repaired with a plastic plate. The study of the microscopic sections dis-closed the tissue to be that of a typical eosinophilic granuloma

Case 6 W O male, age 21 was first seen in 1953 with myo-clonic seizures as his chief complaint. These had occurred since he was 8 years of age. There were no objective or focal signs at this time.

When seen again in 1958 the roentgenograms showed a local area of decreased density 1 x 2 cm in diameter in the left parietal region A central area of increased density was also noted Both tables were eroded A raised tender lesion was palpated in the area involved

It is believed from the history of the seizures, that they are not associated with the local lesion However a pneumoenceph alogram an electroencephalogram and other studies are planned Surgery is to be evaluated according to the findings. This case is listed as a probable eosinophilic granuloma

Postoperative checks have been made on the above patients, and additional roentgenograms have been made of the skull as well but no additional troubles have developed and they remain symptom free



Fig 27 Case 5 Eosinophilic granuloma of parietal area.



Fig 28 Case 6 Eosinophilic granuloma of parietal area.

XANTHOMATOSIS

In 1893 Hand[10] first described xanthomatosis but erroneously associated it with tuberculosis In 1915 Schuller[11] and in 1919 Christian[12] reported some cases of this disturbance but associated it with pituitary dysfunction Rowland[13] in 1928 was first to discover the true nature of the disease, that is, a disturbance of lipoid metabolism Epstein and Lorenz[14] and Cowie and Magee[15] reported more details of metabolic changes. Sosman[16] and Davison[17] suggested that trauma or infection may be causative agents in stimulating local lipoid deposits

It is characteristic of this disease to occur in early childhood and most often in males Exophthalmos, diabetes insipidus and changes in the skull represent the classical symptom triad of the disease. Other bones of the body may be involved The involvement of the retrobulbar area and the hypothalamic area produces the exophthalmus and the water imbalance These changes are reversible however, because after minimal doses of roentgenotherapy normal water balance returns and the eye recedes

The prognosis is good if the ravages of the disease can be held in check until puberty while the reverse is true in those cases starting after puberty or in adult life

Usually both tables of the calvaria are completely eroded The scalp over the defect is elevated but the margins of the bony defect can be easily palpated There is some tenderness The size of the defects vary from a few millimeters to several centimeters in diameter The irregularity of the large lesions accounts for the map-like configuration that is characteristic of this type skull erosion

The microscopic findings are easily recognized and diagnosis is not difficult Histiocytes loaded with fat are found in the peri vascular spaces New connective tissue invasion occurs, with fibrosis and foreign body giant cell reaction Foam cells are found in the marrow spaces of the bone Atrophy and degeneration of the invaded bone develops because of the massive accumulations of foam cells in the marrow spaces It has been thought that fat cells would not invade the bone tissue

Weedmand and Freeman[18] considered nerve tissue as being refractory in lipoid invasion Kyrkland[19] and Henschen[20] found



Fig 29 Case 1 Xanthomatosis showing proptosis.

lipoid changes in the hypothalamus. Deetrick[21] found lipoid tumor masses above the optic nerves Davison[17] found deposit of fat cells in the reparative processes in the cerebrum and cerebellum

We have found 4 cases of xanthomatosis among out records For illustrative purposes, 2 will be included in this study Both were under puberty Small amounts of roentgenotherapy were used over the skull lesions, and over the hypothalamus if exophthalmos and diabetes insipidus were present It was remarkable how much improvement occurred when a minimal dose was used at intervals

CASE REPORTS

Case 1 W R. O age 3 male, was first seen on November 12, 1937 Seven months before, in April the patient had bumped his head with considerable force, so that when a tumor mass was first noticed a few weeks later it was thought that it was a partial detachment of scalp with a hematoma beneath. In July 1937 3 months later protrusion of the left eye was first noted The eye gradually became more prominent A polydipsia and polyuria slowly developed to where the child would drink about 3 times the amount he formerly drank during the day and about 4 or 5 glasses each night, with nocturia 3 to 4 times. In October it was observed that the right leg was weak and there was tenderness on deep pressure over the lower part of the femur He also protected his right upper extremity because of pain in his shoulder



Fig 30a. Case 1 Xanthomatosis, showing map-like configuration of skull defect
Fig 30b Same case showing scapular involvement.

The neurologic history was negative, except for the development of the tumor in the right parietal region, limitation of motion of the right upper and lower extremities and the uni lateral left exophthalmos.

Examination revealed a definite left exophthalmos, a large soft, nonpulsating mass 4 x 6 cm. in the right parietal area and a small bony defect 1 x 1 cm in the right frontal region By careful palpation a discrete bony defect could be made out beneath the tumor mass. Point tenderness was present over the right scapula and over the right femur at the junction of the lower and middle thirds.

Roentgenographic examination showed defects in the skull in the places mentioned also bony defects in the right femur and the right scapula The blood and urinalysis were negative. The blood Kahn was negative The blood cholesterol was 287 mg per 100 cc of serum

The patient was placed on a low fat diet and roentgenotherapy given in small doses, using 200 K.V 20 ma at 50 skin focal distance with filters of 0 5 copper + 1 aluminum. In 2 years, 1 350 r were given in 5 series to the left frontotemporal area 900 r in 5 applications to the right femur and 500 r in 3 times to the right scapula He made a spectacular improvement The large right parietal mass had disappeared 2 months after

350 r had been given and a sunken cranial defect could be palpated in its place. The right upper and lower extremities ceased to cause pain 2 months after 300 r had been given

In 2 years the smaller skull defect was hardly palpable and a large one was less than half the original size and fairly well filled in by fibrous tissue There was no longer a pulsation present Roentgenologically the right scapula and femur appeared normal The left eye was not so prominent as when first seen, but the mother stated that if the patient became excited or had lost sleep the eye protruded more

The polydipsia and polyuria were relieved within 4 months after the first treatment was administered A total of 2 050 r was given to the left temporofrontal and right frontoparietal regions the pituitary and neighborhood area being in direct line of some, it was undoubtedly irradiated sufficiently to produce results. He has grown and developed normally and at present he has graduated from the university is married and has 2 normal children.

Case 2. W L. age 9 was first seen in 1936 In November 1935 4 months previously the patient first noticed a frequency and a gradually increased nocturia which had mounted to 20 times the first night at the hospital He was very thirsty

He complained of a dull right frontal headache, and at times a bitemporal headache He had progressively grown weaker during the past 4 months and weighed only 51 pounds on admission. Since December 1935 3 months before admission, there had been blurring of vision

The child was sluggish and apathetic. The parents stated that he had become mentally dull

On examination the blood pressure was 90/70 The pulse was 70 the temperature normal The lips, hands and feet presented a dusky hue, as if patient were cyanotic. Respirations were normal. The cranial nerves were negative, except for a papilledema of the right disk The pupils were widely dilated and reacted sluggishly to light. There were no other abnormal neurologic findings.

A 1 x 2 cm defect could be palpated in the occipital region, also a 3 x 5 cm defect in the right parietal region.

The specific gravity of the urine was never above 1 005 The red blood cells were 4 730 000 and the white cells 10,200 with polymorphonuclears 65% lymphocytes 32% eosinophils 2%,

monocytes 2% The test for Bence Jones protein was negative. The Wasserman was negative. The sugar tolerance test gave fasting blood sugar of 115 in a half hour it was 220 1 hour 229 2 hours 133 The blood plasma bicarbonate was 40 volumes per cent on February 11 1936 and 50 volumes per cent on March 14 1936 Blood cholesterol was 315 mg per 100 cc. of serum

Vasopressin was given to control the insipidus. The patient voided between 5 000 and 6 000 cc. in 24 hours. The intake was proportionally large With vasopressin the intake and output could be controlled very readily

Roentgenotherapy For a period of 4 days, beginning March 19 1936 407 r were given directly over the lesion in the right parietal area The pituitary area was protected May 1 to May 5 1936 another 444 r were given directly to the pituitary area Two hundred and ninety-six r were given over the cranial defects. April 24 1937 270 r were given over the same area.

It is interesting to note that the diabetes insipidus was not changed in any way after the administration of the roentgenotherapy over the skull defects and that it was necessary to continue with vasopressin However with an application of 444 r or approximately 70% of an erythema dose, directly over the pituitary region, the polyuria with polydipsia gradually diminished and the fluid intake and output returned to normal It has remained normal since, which has been 23 years.

For historical review of the subject, the reader is referred to the reports of Sosman,[16] Davison[17] and Chiari[22]

FIBROUS DYSPLASIA OF THE CALVARIA

Fibrous dysplasia is a disease of young people, seen most often from childhood through the third decade of life. Females are affected more frequently than males Coley[23] suggested that it is a skeletal developmental anomaly characterized by single or multiple areas of fibrous tissue in bone Schlumberger[24] thinks that the single lesion may represent a response to injury to the bone In his report of 67 cases, he noted that the ribs were the most often affected The skull was involved in only 8% of his cases

Lichtenstein[25] noted that the average duration of symptoms in his cases was 20 years This would indicate that the symptoms are not severe and that the lesion is usually not discovered early

in the course of the disease Perhaps in those cases affecting the skull the local enlargement or nodule is noticed more readily than when located on a rib or other well covered bones of the body

There may be a single lesion called a monostotic tumor, or multiple lesions called polyostotic fibrous dysplasia, if more than one bone is involved The former is more common The latter is often associated with other systemic changes, such as skin pigmentation and precocious sex development

In 1937 Albright[24] and associates were first to describe as an entity the disturbance now known as ' Albright s Brown-spot Syndrome, or disseminated fibrous dysplasia with cutaneous manifestation and sexual precocity The skull is often involved in this type of case, and presents a problem secondary to systemic involvement elsewhere We will not further elaborate on this type of disturbance

In some cases of polyostotic types, and in most cases of monostotic types, and particularly where the skull alone is involved there may be 1 or more lesions but there is not an associated systemic disturbance with skin discoloration or sexual precocity There may be 2 or more lesions in the same bone and still be a monostotic lesion A polyostotic lesion refers to lesions involving more than 1 bone The lesions may be of variable size, depending on how early it is discovered Some soreness is present over the raised area and there is tenderness on pressure.

Roentgenograms show a rarefied well demarcated radiolucent lesion associated with thickening and sclerosis in places, while at other areas, erosion may be noted A multilocular appearance, as if cystic, is often seen and at times it is difficult to differentiate it from a cystic tumor If the sclerotic changes predominate, it is sometimes difficult to rule out a bony tumor A biopsy may be required in some of the solitary lesions in order to make a diagnosis We think that at this time, if the frozen section proves the lesion to be fibrous dysplasia, *that one should be prepared to* remove the entire lesion and mold a plastic plate into the defect. According to Coley [25] there is a likelihood of transformation to a sarcoma Notwithstanding the tendency for a progressive enlargement associated with pain, would support the wisdom of removal

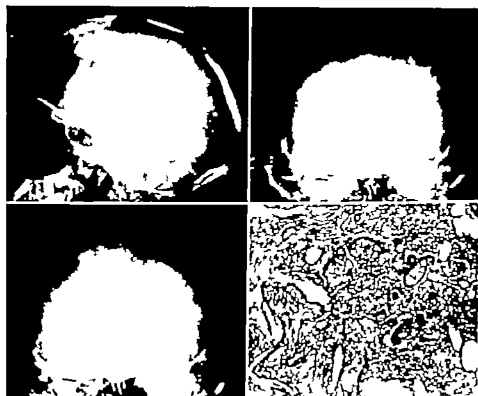


Fig 31a (*Upper left*) Typical appearance of fibrous dysplasia of calvaria
Fig 31b (*Upper right*) Same case, p.a. view
Fig 31c. (*Lower left*) Same case operative defects.
Fig 31d (*Lower right*) Same case, photomicrograph low power

The microscopic findings of the lesion without, and with the associated skin and endocrine changes, differ considerably So much so that trauma is suspected as the origin of the former and a congenital origin for the latter Schlumberger[14] was unable to find islands of cartilage in the primary tissue of any of the 67 cases of monostotic fibrous dysplasia that he had studied He supported the theory of injury as a causative factor The fibrous tissue cells were uniform and well differentiated with small spicules of bone interspersed throughout the field

At surgery it is best to plan the removal of the entire lesion by curet and rongeurs Sometimes this enlarges the original bony defect considerably but this removal must be extended to good bone about the entire margin of the lesion Some places the

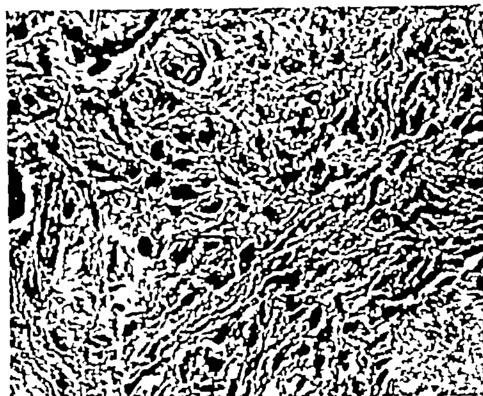


Fig 31c Same case, photomicrograph, high power

tissue may be quite soft It is very vascular A plastic repair of the skull permits the use of any size or shape needed and lends itself well to these cases

CASE REPORTS

We have only encountered 3 cases of fibrous dysplasia of the skull

Case 1 B H was a female, age 17 whom we saw in 1954 because of 2 knots, 1 in each frontal area The 1 in the left frontal area was tender and was about 4 x 5 cm. in diameter The rightsided lesion was approximately 2 x 3 cm in diameter This was a questionable monostotic type because of the slight extension of the leftsided lesion back into the parietal bone However it seems to have originated in the frontal bone bilaterally thereby qualifying for a monostotic type Besides, roentgenograms of the entire skeletal system did not reveal any other lesions.



Fig 32. Case 2. Monostotic fibrous dysplasia

In January 1954 through a coronal incision, both lesions were removed completely and a plastic plate molded into the defects. The scalp and dura were not involved The microscopic studies revealed mature fibrous tissue proliferation with spicules of bone interspersed No cartilage was found There was no evidence of malignancy

Subsequently in 1956 and 1957 roentgenograms showed no change in the skull or skull margins about the 2 cranioplastic repairs The plastic plate permits good visualization of the skull margins. There were no other complaints.

Case 2. P H a female, age 17 was referred in 1950 because of a lump on the right side of the head, and a dull ache over the entire head About 3 years before, when she first noticed the lesion, it was about the size of a dime. When examined in the right parietal area there was an area, 4 x 5 cm of decreased density also there was some bony proliferation The defect was irregular and involved both tables and reported as a fibrous

dysplasia Roentgenograms of the pelvis, extremities and chest with ribs, were negative. At surgery a square bone flap 4 5 cm each way was turned down about the lesion The center of the flap corresponding to the tumor measured 2 cm in thickness A tantalum plate was fitted and secured with 3 screws

The histologic sections showed considerable fibrous replacement and proliferation in the cancellous portion of the bone Dr Black reported further that there were many thick bony trabeculae running through this very thick fibrous tissue. There were a number of multinucleated giant cells present and many of these appeared as the epulis type giant cell His diagnosis was "fibrous dysplasia of bone.

This patient has been observed at yearly intervals. There have been no other troubles. Recheck of the skeletal system remained negative. She has since married

Case 3 J F R male age 20 was seen in 1958 because of a tumor over the left eye first noticed after being struck with a brick 12 years previously It had slowly grown larger until now there was noticed a bulging area in the left frontal region, extending from the lateral margin of the frontal sinus to the lateral confines of the skull and apparently into the tempor ozygomatic bones A nodule about the size of the end of his index finger protruded from the lateral margin of the orbital ridge, displacing the left eye slightly mesialward He had had headaches on the left side, and had had some dizziness, nausea and vomiting There had been no report of double vision up to date.

Roentgenograms showed an area of increased density with altered texture in the bone forming the lateral margin of the left orbit The zygomatic process and orbital plate were also involved A tangential view showed a localized bulge in the frontal bone, which was evidently due to an overgrowth of rather thinly trabeculated bone. In the area of the bulge the tables were thin. The outer table was expanded and the diploic space was widened with an altered texture. Medially the disturbance extended to the margin of the frontal sinus. The superior orbital rim was flattened and there was slight downward encroachment upon the space of the orbit. There was increased prominence of the lateral margin of the base of the zygomatic process. The roentgenologist's diagnosis was that of a variant fibrous dysplasia

At surgery the left frontotemporal bone was exposed with a flap turned at the base over the supraorbital ridge. The supra



Fig 33a. (*Upper left*) Case 3 Photo showing fibrous dysplasia of left supraorbital area.
Fig 33b (*Upper right*) Same case, showing lateral encroachment upon left orbit.
Fig 33c (*Lower left*) Same case, roentgenogram showing involvement supraorbital area.
Fig 33d (*Lower right*) Same case roentgenogram showing lat. view

orbital ridge was exposed A definite bulge and thickening of the bone was visible. Three burr holes were placed in the superior lateral margin of the exposure at the periphery of the tumor after which rongeurs were used to remove the bony tumor

In places the tumor was 3 cms. in thickness. The 2 tables of the bone were obliterated by the intervening tumor tissue,



Fig 33e. Same case showing postoperative skull defect (p.a view) and wires securing plate in place

which seemed to have grown into and through them The tumor was rather soft, considering that it was bony and could be removed easily with rongeurs without much force. It was more vascular than normal bone

The entire tumor was removed to good bone around the periphery This necessitated removal of part of the frontal bone outer two-thirds of the supraorbital ridge, the orbital plate part of the zygoma and part of the temporal bone There was no soft tumor tissue encountered

A plastic plate was molded and fitted in place and secured with 3 stainless steel wire sutures. It was possible to mold a new supraorbital ridge and to cover over the orbital cavity

The specimen was decalcified and sectioned The microscopic sections revealed the tumor to be a fibrous dysplasia of the calvaria. This is the largest lesion of this kind that we have encountered It was totally removed He should have no more trouble from this lesion

PAGET'S DISEASE

Osteitis deformans was first described by Paget[37] in 1877, as a form of chronic inflammation of bone. Inflammation is still considered a likely cause by many Endocrine dysfunction is also supported as an etiologic factor Actually, a proven cause has not been definitely established The etiology is unknown

It is most always multicentric, affecting most bones of the body, except rarely the hands and feet. It involves males in an overwhelming majority and the skull in a high percentage of cases. Bird[18] estimated that Paget's Disease occurred in 1 3000 patients admitted to the hospital This number may be high Certainly the skull involvement is considerably less frequent

The onset is so insidious as to be imperceptible, and when first noticed may have been quietly developing for years At this stage, and it is usually in a male past middle life one finds a rather limited area or areas of decreased density of osteolytic type involving the outer table of the calvaria Ritvo[19] emphasizes the presence of a narrow band of slightly increased radiance at the peripheral margins of the diffuse rarefaction, as a diagnostic feature of early Paget's Disease At this time the bone is soft and shows no sclerosis. Later a poor quality of new bone is formed involving the outer table and diploe first then the inner table as the lesion advances. It becomes hard and takes on a ' cotton tuft appearance The skull becomes enlarged asymmetrically The features are distorted There may be pain of moderate degree. No temperature elevation is present Serum phosphatase may be elevated Bird[18] found that 11% of cases studied, developed osteogenic sarcoma Other authors report similar findings.

The microscopic findings vary somewhat, depending on the *stage of the disease Early the tissue is soft,* vascular and fibrotic Osteoclasts are present Huge deposits of bone tissue develop with irregularly placed trabeculae. Osteoblasts are more numerous in this stage

Surgery is not indicated. The foramina are not encroached upon, causing little involvement of the nerves and blood vessels of the area involved. Roentgenotherapy has been used for palliation of pain. Continued observation is essential to apprehend the development of osteogenic sarcoma. Symptomatic treatment is otherwise indicated.

CASE REPORT

We have encountered only 1 case involving the skull. It may be that this lesion is readily recognized by the profession and is not referred because surgery is not indicated and treatment is limited. Notwithstanding, in the thousands of skull films obtained for other reasons during the past 27 years we have not inadvertently discovered more cases of Paget's Disease of the skull.

C. H., male, age 61, was referred in 1945 with a diagnosis of Paget's Disease with a request of his family physician for any suggestions that may prolong the patient's active career. He only recently discontinued work as a plumber because of confusion, some headache and difficulty in getting about. Ten years before he had fallen. He had bumped his head but was not unconscious. The size of his hat had increased from 7¼ to 8¼ in the past 10 years.

There was no increased intracranial pressure and no focal signs found. Aside from mild confusion and headache, neurologic studies gave normal response. Extensive laboratory investigation showed well developed Paget's Disease, with typical roentgenographic findings.

The patient was treated symptomatically and expectantly by his family physician.

LEONTIASIS OSSEA

There is considerable difference of opinion concerning this disease. Ritvo[19] thinks it is not an entity but that various types are caused by different etiological factors. Leontiasis is a descriptive term that has been used for centuries applied to various conditions. Virchow,[20] however, in 1865 used it to describe a disturbance of a bony type involving the face.

Leontiasis ossea is a rare condition beginning around puberty and confined to the bones of the head. According to Reisz,[21] the

bones of the upper part of the face and the frontotemporal area are more frequently involved It is slowly progressive Gradually the physiognomy takes on the appearance of a lion The foramina and canals of the skull are often obliterated, resulting in auditory visual lacrimal and nasal disturbances of varying degrees, depending upon the stage of the disease Sometimes the encroachment upon the orbital fossa may produce proptosis and upon the optic foramina, blindness

The proliferation of bone may be quite extensive, at times becoming several centimeters in thickness The roentgenograms reveal the involvement of the upper bones of the face and the frontotemporoparietal area mainly There may be massive production of dense bone with a loss of definition of the skull tables There is a periosteal production of sclerotic bone with overproduction of osteoblasts

No definite etiologic factor has been discovered Treatment is mostly symptomatic In one of our cases with marked proliferation of bone and pressure symptoms involving the frontal area and producing proptosis surgery was used to obtain mechanical benefits and to relieve the effects of pressure

CASE REPORTS

Case 1 R H female, age 55 was referred in 1945 because of the enlargement of the face and skull in the frontal area first noticed about 25 years previously Protrusion of the left eye had been developing for 5 years The furrowed features were typical of leontiasis ossea. Also the massive enlargement of the frontotemporoparietal bones with associated involvement of upper facial bones and a left proptosis, indicated a far advanced case *In places, the bone growth was over 5 cm. in thickness.* Headache and visual disturbance (blurring) were becoming more troublesome

After a considerable amount of planning surgery was advised with the idea of reducing the blood supply to the area involved and decompressing the left eye February 28 1946 under basal local anesthesia the left posterior margin of the tumor was exposed Because of the vascularity and dural adhesions, a guide could not be used Instead a rongeur was used to remove a section of bone at the periphery extending from burr hole to

burr hole Bone wax in large quantities was used to control bleeding When we found a gigli saw was not usable we tried an electric rotary saw Bleeding could not be controlled satisfactorily so it was abandoned When a strip of bone 2 x 30 cm had been removed we placed tantalum foil between the margins and closed the scalp tightly over it A pressure bandage was used Seven days later the procedure was continued The small frontal sinus was uncovered and the membrane peeled away uninjured Then, with multiple burr holes directed through the lower frontal bone and downward the thickened skull was penetrated into both the frontal fossa and orbital fossa With rongeurs it was possible to enlarge the burr openings so that the thickened orbital roof could be removed as well as a nubbin of bony tumor extending into the retro-orbital space The retro-orbital fat protruded into the decompression

With rongeurs and burr the trench about the tumor was continued about its entire circumference connecting on both sides with that made the previous week This left only a bridge of bony connection from the tumor mass by way of the left malar area to the lesser wing of the sphenoid A good bit of this bridge was removed but not all of it However the vascular connection was otherwise completely severed

The dura was divided all around except for the sagittal sinus. The middle meningeal arteries were ligated At this point it was decided that the tumor and involved calvaria could not be safely removed Thinking that the blood supply was materially reduced and the eye decompressed we proceeded to remove the external mass of the tumor by making multiple perforator and burr openings parallel to the inner table and removing the intervening bone with rongeurs. This gave a good cosmetic result

Subsequently the eye receded from its place of prominence, headaches were relieved and the skull enlargement was less prominent In the time that has elapsed approximately 12 years, there has been no apparent evidence of growth, recurrent proptosis or headache.

Case 2 S A S female, age 16 referred in 1957 because of unsteadiness of gait and myoclonic seizures. It was noticed that the left maxilla and zygomatic area was considerably enlarged but when inquiry about this was made, the family indicated that this had been present since early childhood and although it was becoming slowly larger they were concerned about the seizures



Fig 34 Leontiasis ossea, showing thickening of the maxillary zygomatic and temporal bones.

and unsteadiness. The seizures began 6 years before and lately some unsteadiness and personality changes were noticed

Aside from the history and appearance of the face, we could find no abnormal objective findings on complete physical and neurologic study The roentgenograms taken in 1957 showed some increased density and thickening of the temporo-zygomatic malar area as compared with some taken in 1953 Pneumoencephalograms showed no displacement or impingement upon the ventricles. The cortical markings were normal. Spinal fluid studies were normal

It was possible to control her seizures by changing her medication and with this her personality and unsteadiness improved

HYPEROSTOSIS FRONTALIS INTERNA

In 1936, Moore[22] described a hyperostosis frontalis interna in a paper on metabolic craniopathy Since it has often been referred to as Sherwood Moore's Disease Its cause is unknown

The disease is not uncommon The hyperostosis is associated with or without symptoms Usually it occurs in females between the ages of 35 to 45 What the connection may be between the hyperostosis, a dull headache personality changes, obesity and other mild metabolic changes has not been determined Many cases have no symptoms associated with the hyperostosis

The roentgenograms show a bilateral symmetrical dense, irregular wavy proliferation of bone involving the inner table only and projecting inwardly The diploe is not involved There is some calcification of the falx and at times along the walls of the sagittal sinus Surgical treatment is not indicated but symptomatic treatment may be helpful

We have on record a large number of cases of hyperostosis frontalis interna The findings and symptoms conform so closely that it seems unnecessary to relate the case record on all so we have chosen at random 4 cases for brief presentation

CASE REPORTS

Case 1 J B H age 55 referred in 1949 because of a dull aching and compression feeling in the vertex of the head. There was some insomnia and tension problems. She was moderately obese Complete physical and neurologic studies revealed no organic changes of abnormal character Roentgenograms showed rather marked internal hyperostosis bilaterally in the frontal area Her headaches gradually subsided over a period of years, and although there is some discomfort, she understands her condition and has learned to live with it.

Case 2 E. S female, age 57 referred in 1951 because of a headache and a feeling of fullness, beginning in her early 40s. The only objective finding on physical and neurologic examination, was a right nerve deafness. Encephalogram showed normal ventricles and subarachnoid space Spinal fluid findings were all normal Roentgenograms revealed an advanced hyperostosis frontalis interna normal internal auditory meati and petrous ridges. She made a good adjustment to her problems, after she was reassured that she had nothing to fear

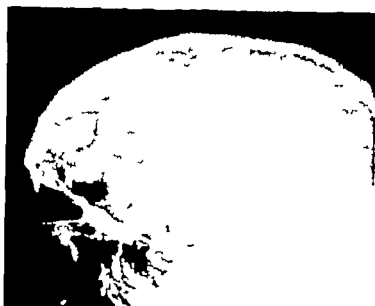




Fig 35a. Case 3 Hyperostosis frontalis interna lat view
Fig 35b Same case, p.a. view showing calcification of falx.



Fig 36 Case 4 Hyperostosis frontalis interna

Case 3 S K female, age 46 was referred in 1955 because of frontal headaches, and some feeling of unsteadiness. These started after a car accident 3 months previously There was a scar in the right supraorbital area where she had been struck but not rendered unconscious. Roentgenograms showed a typical picture of hyperostosis frontalis interna with bilateral bony proliferation and calcium deposits in the falx It was thought from her history that this lesion was asymptomatic and that her headaches originated from the head injury Subsequently her headaches left her

Case 4 S R female, age 44 was referred in 1950 because of a continuous headache and a feeling of weakness. The neurologic examination did not reveal increased intracranial pressure or focal signs or symptoms. The sella was slightly enlarged There were no neighborhood symptoms Visual fields were normal. Glucose tolerance test was normal There was also a

bilateral hyperostosis frontalis interna and a calcification of the falx. This patient has been treated symptomatically and kept under observation Headaches have subsided

BIBLIOGRAPHY

1 Otani G Ehrlick J C. Solitary granuloma of bone simulating primary neoplasm *Am J Path 16* 479-490 July 1940

2. Green W T Farber S Eosinophilic or solitary granuloma of bone. *J Bone & Joint Surg 24* 499 526 1942

3 Thaunhauser S J Eosinophilic granuloma of bone *J A M A 134* 1437 1438 1947

4 Swain G Williams, J M Eosinophilic granuloma of bone *J Neurosurg*, *10* 185 1953

5 Knighton, R S Fox J deW Diagnosis and treatment of eosinophilic granuloma of skull *J A M A*, *162* 1294-1297 December 1956

6 Shuster B H Flynn, T F Eosinophilic granuloma of frontal bones report of case. *Arch. Otolaryng 60* 501 504 October 1954

7 Raaf J Eosinophilic granuloma of the skull *West J Surg* p 53 Jan 1945

8 McNeil, Clyde Personal communication.

9 Baker G S Fisher R. G Eosinophilic granuloma of the skull report of 4 cases with 5 year follow up N.S N.D

10 Hand A General tuberculosis *Trans Path Soc Philadelphia*, *12* 282 284 1893

11 Schuller A. Ueber ergenartige Schadeldefekte un Jungendolter *Forsch a. Geb Rontgenstrahlen*, *23* 12 18 1915

12 Christian, H A Defects in membranous bones, exophthalmos and diabetes insipidus An unusual syndrome of dyspituitarium a clinical study in *Cont Med & Biol Research* Vol 1 pp 391-401 New York, Hoeber 1919

13 Rowland R. S Exanthomatous and reticulo-endothelial system Correlation of unidentified groups of cases described as defects in membranous bones, Exophthalmos and diabetes insipidus (Christian s syndrome) *Arch Int Med 42* 611-674 1928

14 Epstein Emil Lorenz, Karl *Ztschr f Physiol Chem 192* 145 1930

15 Cowie, D Magee, M C. Lipoids and lipoid diseases Disease (Xanthomatous) and review of the literature on the lipoid content in human tissue *Arch Int Med 53* 391 March, 1934

16 Sosman M Xanthomatosis. *J.A.M.A 98* 110 January 1932

17 Davison C. Xanthomatosis and the Central Nervous System *Arch Neurol & Psychiat* *30* 75 July 1931
18 Weidman F Freeman W Xanthoma Tuberosum *Arch Dermat & Syph* *9* 149 February 1924
19 Kyrklund R Beitrag zu einem seltenen Symptom-Komplex *Ztschr Kinderh* *41* 56 1926
20 Henschen F Fall von Xanthomatose mit Knochenveranderungen und grosszelliger Sphenomegalie *Zentral bl allg Path.*, *50* 44 1930
21 Dietrich A *Virchows Arch Path Anat* *212* 119 1913
22 Chiari, H Die generalisierte Xanthomatose von Typus Schuller Christian *Ergebn allg Path* *24* 396 1931
23 Coley B L. *Neoplasma of Bone* New York Hoeber 1949
24 Schlumberger H G Fibrous dysplasia of single bones (monostotic fibrous dysplasia) *Mil Surgeon* *99* 504 1946
25 Lichtenstein, L. Jaffe H L. Fibrous dysplasia of bone. *Arch Path* *33* 777 1942.
26 Albright, F Butler A M Hampton, A O Smith, P Syndrome characterized by osteitis fibrosa disseminata areas of pigmentation and endocrine dysfunction, with precocious puberty in females report of 5 cases. *New Eng J Med* *216* 727 746 April 29 1937
27 Paget, Sir J On a form of chronic inflammation of bones (osteitis deformans) *Tr Roy Med Chir Soc London*, *60* 37 *65* 225 1877 and 1882
28 Bird, C. E. Sarcoma complicating Paget s disease of bone report of 9 cases, 5 with patholigic verification *Arch Surg* *14* 1187 1927
29 Ritvo Max *Roentgenographic Diagnosis of Diseases of the Skull* New York, Hoebner 1949
30 Virchow R. *Arch Path Anat* 1865
31 Reisz M Leontiasis ossea. *Arch Klin Chir* *184* 320 1936
32. Moore, Sherwood Metabolic craniopathy *Am J Roentgenol* *35* 30-39 1936

Chapter IV

MALIGNANT TUMORS OF THE CALVARIA

We have included in this group 8 tumors definitely malignant and which have a tendency to develop as primary tumors of the calvaria (see *Tumor Classification* p 3) There are others that may be included such as, Hodgkin s disease and lymphosarcoma but have been left out, either because they are encountered so infrequently in the calvaria or their presence in the skull does not constitute a part of a symptom complex of that particular general disease or the customary primary site of origin

OSTEOGENIC SARCOMA

Coley[1] found only 7 cases involving the skull in the files of the Memorial Hospital over a period of 20 years Three were in the parietal bone, the others in the basilar bones of the skull Geschikter and Copeland[2] also found the calvaria to be quite rarely involved by osteogenic sarcomas Except for the associated development in Paget s disease, they tend to occur in the younger individuals

The patient complains of pain and a swelling of the head Both rapidly progress Early metastases to the lungs soon cause additional symptoms of a systemic character

Roentgenograms show marked bone destruction with new bone formation The growth extends from the skull both internally and externally Fortunately however it seems to develop outwardly more rapidly Its extension through the pericranium into the scalp produces at times a sizable tumor and considerable pain There is not a comparable inward extension, and, although the dura may be involved and compressed inwardly it is not likely to be penetrated

At surgery a varying degree of vascularity is found in different parts of the tumor Also a varying degree of ossification is found

Some areas may require rongeurs for removal, while a curet will do quite well in other places In one patient there were 2 distinct separate elevated areas in the frontal bone that represented tumor tissue that was densely ossified while anteriorly where it extended onto the orbital plate and lesser wing of the sphenoid bone, the tumor tissue was softer and could be easily scraped out with a curet It was perhaps 1 to 1 1/2 inches in thickness and quite vascular

These patients do not live long Surgery may help prolong life and certainly can reduce suffering Cade[3] applies roentgenotherapy to the lesion and observes them for 2 or 3 months after which he operates only those in whom no metastases have been found

They seem to get some relief after the tumor has been removed As an additional helpful measure we recommend that at surgery a plan also be made to section the posterior root of the 5th nerve on the same side, if the lesion is frontal and basilar The removal of the tumor relieves the pain incident to the invasion of the pericranium and the scalp as well as that caused by pressure from expansion, while the posterior root section will anesthetize the area supplied by the 5th nerve. It is helpful In time, other pain relieving devices and medication may be needed Eventually and not too long afterwards they succumb with metastases to the lungs and other bones of the body

CASE REPORTS

Case 1 R. S male age 15 years, was referred in November 1957 because of a proptosis of the right eye, a mass in the right frontal area and headache. These symptoms had begun about 3 months previously at which time he had fallen off a bicycle injuring this area

At 15 years he was 6 feet 2 inches tall and weighed 182 pounds. BP 150/100 General physical examination showed an abnormally large boy and rather obese, with normal findings, except for those factors relative to the right frontal area There was no increased pressure or focal neurologic signs. The eye was proptosed and displaced mesialward and upward There were 2 distinct and separate masses, about the size of a lime in the right frontal area

Roentgenograms showed the 2 areas to be somewhat denser than the surrounding skull Both were elevated above the outer table. There were some regions of decreased density There was thickening and increased density of the zygoma, the orbital roof and the lesser wing of the sphenoid It was evident that we were dealing with a single tumor presenting varying degrees of density bone proliferation and destruction.

At surgery the involved frontal bone and elevated masses were removed with rongeurs. Then the orbital plate, parts of the zygoma temporal bone and part of the lesser wing of the sphenoid bone were removed with rongeurs and curet. Some of this tissue was quite soft and some quite hard Bleeding varied considerably throughout various areas of the tumor

The pathological report indicated there were about 70 gm. of tumor sent to the laboratory Large areas of the tumor were composed of fibrous type tissue. The nuclei were irregular varying moderately in size and staining properties. A moderate amount of mitosis was observed. There were many areas in which the tumor was osteoid and bone was present which had the appearance of being tumor bone. Cartilage cells were also present in some portions of the tumor Other areas showed fibroblastic tissue, which appeared to be directly producing osteoid Diagnosis was osteogenic sarcoma.

Subsequently the patient continued in school. His eye had returned to its normal position He was contented and felt well. In April, about 4 months later pain became a matter of concern. It was mostly in the maxillary distribution At this point we wished that a posterior root section had been done at the time of the tumor surgery because it would have saved him another operation. This pain became so acute that the patient readily agreed to have the posterior root of the 5th nerve sectioned. This again gave him relief for 4 more months. The subsequent section of the posterior root has certainly been worthwhile, even though a bilateral mesial frontal lobe section has since been necessary to control an apprehensive and emotional state.

An enlargement of the right side of the face was quite noticeable when last seen, and it is evident that the tumor has continued its growth in the base of the skull and face. The zygomatic process, the squamous portion of the temporal bone, and the malar bone have recently become involved Pressure over the malar enlargement does not cause pain. There has been no noticeable

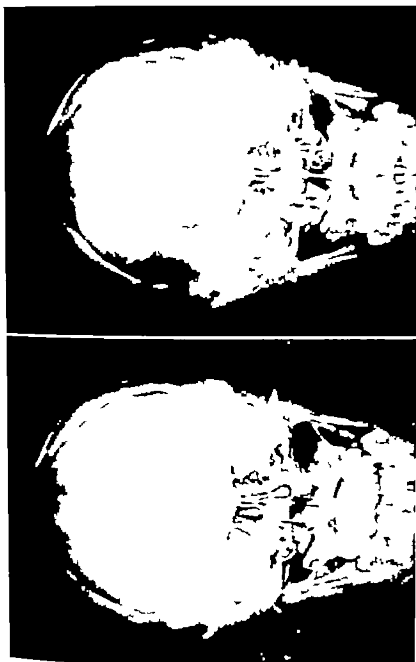


Fig 37a Case 1 Osteogenic sarcoma Involving frontal bone zygoma orbital plate and sphenoid bone
Fig 37b Same case showing postoperative defect in skull.

enlargement or evidence of recurrence of the tumor along the outlines of the skull defect The craniectomy opening has been a little sunken and soft and has shown no signs of intracranial pressure to date (September 1958)

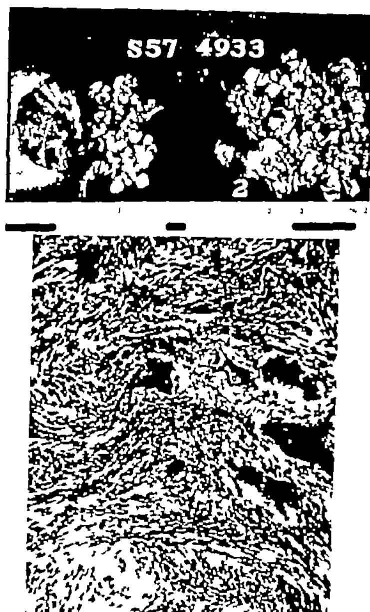


Fig 37c. Same case, specimen showing dense ossified tissue to left and softer tissue to right

Fig 37d Same case, photomicrographs of osteogenic sarcoma, showing tumor bone formation

Case 2 C P male, age 14 years, was referred in May 1948 because of a large tumor on the right side of the head posteriorly During the past 5 months the lesion had been operated twice in



Fig 38 Case 2. Rapidly growing osteogenic sarcoma

different cities. It had again attained the size of a flattened grape fruit. This was estimated to be about 10 times larger than the original tumor removed 5 months before and 4 times larger than the tumor removed 2½ months previous to our contact with the patient.

The patient was emaciated cachetic and showed evidence of a chronic illness. There was a secondary anemia Roentgenograms of the chest did not reveal a metastasis and on general examination none was found elsewhere The lesion did not seem to produce intracranial pressure and there were no focal neurologic signs.

Roentgenograms of the head showed a large radiopaque mass in the right parieto-occipital area The protruding tumor contained a large amount of osseous tissue with radiating spicules extending perpendicular to the skull The skull was involved but the tumor did not appear to extend into the cranial cavity There

was a triangular defect in the tumor mass, which appeared to be due to previous surgical removal of bone

At this time, 2,200 r were given through each of 2 portals. For 2 months he improved. In July 1 week before he died he had difficulty swallowing and vomited frequently which may have been due to his cachetic state, or a terminal development of intercranial pressure.

Excerpts of postmortem findings There is evidence of marked cachexia, emaciation and dehydration. A mass 15 cm. in diameter protruded from the right parieto-occipital area There are no lymph gland enlargements noticeable. No fluid in peritoneal or pleural cavities. Pericardial cavity normal Gross examination reveals many calcified lesions near the periphery in both lungs. They range up to 1 cm in diameter Liver weighs 800 gm. The heart, kidneys, G I trace, pancreas and adrenals did not show gross or microscopic evidence of metastases. Mediastinum and retroperitoneal spaces were searched and no lymph node enlargement found Permission was not granted for a head examination.

Microscopic examination of the nodes and viscera did not reveal metastases anywhere, except the lungs. It was necessary to decalcify the lesions in the lung before sectioning They presented neoplastic tissue consisting of many irregular cells with varied shaped hyperchromatic nuclei These, in many instances, produced definite osteoid tissue

CHONDROSARCOMA

Chondrosarcoma of the skull is a very rare tumor It occurs more frequently in the basal area, and almost never in the calvaria as a primary tumor Secondarily it may complicate Paget's disease, or arise from a chrondroma, which in the vault, occurs very infrequently (see p 26 Chondromas) This lesion tends to occur in adults, grows more slowly as a whole, than osteogenic sarcoma Yet it is almost invariably fatal Sometimes surgery will prolong life Irradiation is used separately or with surgery Both give little more than temporary relief In Krantz's[4] case both were used and the patient died 20 months later because of massive intracranial extension of the tumor

Ordinarily metastases or spread occurs by extension along eroded venous channels, or by the blood stream. The heart and lungs are the most common sites of metastases

The experiences we had with the case reported below were quite unique The consensus of opinion was that the tumor was a chondrosarcoma yet this opinion was not unanimous by any means

In this case the manner of involvement of the various areas affected was quite difficult to explain satisfactorily The idea of multicentric development was advanced but the matter is still open to question

CASE REPORT

We have had only 1 somewhat debatable case of chondrosarcoma of the calvaria The case history roentgenograms and tissue sections were circulated by Dr Black pathologist among several eminent authorities on bone tumors, and it was the consensus of opinion that the tumor was a chondrosarcoma of multi centric origin

The patient was a male 26 years of age He was first seen in March 1954 because of constant aching during the previous 6 months, in the left axillary region. Complete studies by an internist gave normal findings, including chest films, electrocardiograms, pyelograms, serum phosphatase, etc There was no focal central nervous system involvement, and no evidence of peripheral motor or sensory changes, except the subjective symptoms of pain

When again examined on July 1 1954 slight motor weakness was found in the left lower extremity There was an indefinite level of hypalgesia at T 7 on the right. There was an associated temperature loss, also Deep reflexes were slightly increased in the left lower extremity Babinski, Gordon and Oppenheim were absent. Laminagraphs showed a tumor in the posterior mediastinum and slightly to the left. The 5th intervertebral foramen was enlarged and there was evidence of destruction of bone tissue

On July 9 1954 in association with a thoracic surgeon, the chest was opened on the left side by removal of the 5th rib and extending the incision far posteriorly

A tumor nodule the size of a lime, was found at the 5th inter vertebral foramen, and another the size of a hazelnut beneath it The transverse processes, the pedicles and portion of the laminae of the 5th and 6th vertebrae were removed The bone was invaded by the tumor In places it was soft and quite vascular After exposure was completed the tumors were removed The dura had not been invaded All abnormal looking soft and bone

tissue was removed This finally included part of the posterior and lateral portions of the body of the 5th thoracic vertebra.

Symptomatically the patient did well. He was free of pain, the motor and sensory functions returned to normal. He continued to work and had few complaints. Repeat roentgenograms of the thoracic spine 6 months later showed bone regeneration in T 5 and T-6 vertebrae.

Subsequently roentgenograms were taken of the entire skeletal system. Poorly defined irregular areas of decreased density were found in midparietal and right parieto-occipital region A radiolucent destructive process involved the manubrium. There was a vague, poorly defined area of decreased density in the right side of the sacrum The cervical, thoracic and lumbar spine were negative, except for the previous findings of bone regeneration at site of the removal of the tumor at T 5 and T-6 The lungs were not involved The 5th rib had partly regenerated. The long bones were negative.

In April 1955 the 2 lesions of the skull were exposed. Soft tumor tissue was found under the pericranium and extended through the skull to the dura, where it spread out between the dura and skull. The involved skull and tumor tissue were removed Again, the tumor was very vascular

The pain and headache were relieved after the removal of the tumors of the skull, and did not return throughout the remainder of his disease, although before he expired, a little over a year later the cranial tumors had reappeared

In October 1955 tenderness was noted over the right sacroiliac region, and roentgenograms showed a small area of destruction in the right sacroiliac area A few weeks later symptoms of cord involvement at T-4 level developed Tumor was removed from the right sacroiliac region and through a laminectomy an extradural mass removed at T-4 level Again, the tumor was very vascular and again the microscopic findings were all similar to the tumor removed before.

The original tumor tissue was thoroughly studied as well as the subsequent specimens. It was after considerable pathologic consultation, and after exchange of clinical records, roentgenograms, as well as sections, that a final diagnosis of a multicentric chondrosarcoma was made. We are deeply indebted to the pathology department for their labors and helpful information

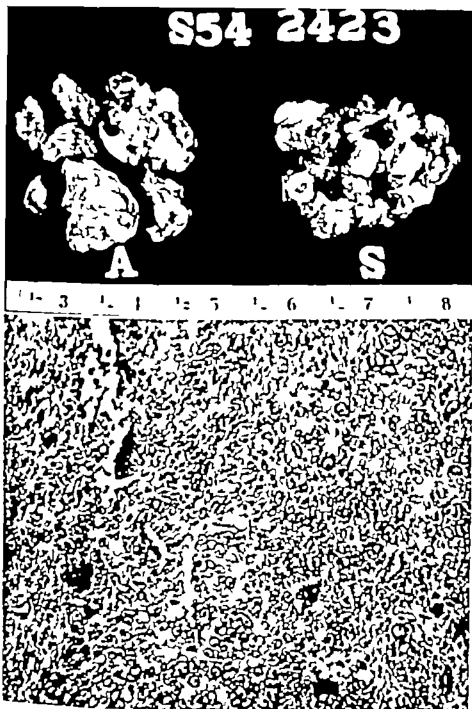


Fig. 39a. Unusual multicentric chondrosarcoma, A. tumor from intrapleural extension S extradural portion of tumor
Fig 39b Same case showing tumor cartilage.



Fig 39c. Same case, showing pattern of undifferentiated neoplasm with areas of tumor cartilage

The tumor was friable and pinkish gray in color A number of fragments of bone were scattered throughout the soft tissue. The pathologist further describes other portions of the tumor as being quite firm and containing tissue, which is either cartilage or very dense collagenous tissue containing some small fragments of bone. Microscopically the report on the skull tumors was in part as follows *The tumor cells range from rather immature* fibrous tissue type cells up to cells that have the appearance of mature cartilage. The cells are invading bone and some spicules of degenerated bone are present within the tumor masses He commented further The neoplasm in this case is unique and some, who have studied the histologic sections, are uncertain as to the proper classification. At first the tumor removed at T 5 area was thought to be probably benign, but it is now obvious that the tumor is malignant. Sections, roentgenograms and clin ical data have been submitted to various pathologists and emin

ent authorities on bone pathology Unclassified tumor, malignant, and chondrosarcoma of multicentric origin in the skeleton received the most consideration after a long and arduous effort to identify the tumor

Roentgenotherapy was used over the local tumor areas, later when recurrence occured It gave some relief of pain This patient remained active for almost another year During the last 3 to 4 weeks he stayed indoors and in bed most of the time There was never evidence of pulmonary or other visceral involvement He succumbed to the effects of the tumor October 1956

FIBROSARCOMA

According to Pancoast, Pendergrass and Schaeffer,[3] fibrosarcomas do involve the cranium This supports the concept of Geschickter[6] that 'they arise from the outer layer of the periosteum or from the surrounding soft parts Lichtenstein[7] mentions a type of fibrosarcoma that originates centrally It is primary in the medullary region

The above authors do not report a case involving the calvaria and I have been unable to find reference of a case Kirschbaum[8] in 1943 reported a case associated with Paget s disease of the skull A primary fibroma of the calvaria was reported by Dandy [9]

EWING S SARCOMA

Coley[1] states that Lucke in 1866 was first to recognize this tumor It has been known as round cell sarcoma In 1921 Ewing[10] described in detail the various aspects of this lesion, and since, it has been referred to as Ewing s tumor Next to osteogenic sarcoma Ewing s tumor is the most common primary tumor of bone Yet it is almost never found as a primary tumor of the skull It may metastasize to the skull If one is found here a primary lesion elsewhere is most certainly to exist. Since it does occur so frequently it is thought best to mention it, in spite of its tendency to almost always develop in other places than in the calvaria

It develops most frequently in male children, in the shaft of some long bone Although it is very sensitive to roentgenotherapy the prognosis is not good It is considered advisable to secure

a biopsy before roentgenotherapy is used, so as to allow a microscopic study of more nearly natural tumor tissue Again if found in the calvaria it is most likely metastatic

MYELOMA (PLASMA CELL)

Myeloma is a disease of later life affecting males more frequently It is generally accepted that it originates from the blood forming elements. There is some variation of opinion as to whether it should be classified as a neoplastic disease Certainly, myeloma tissue develops in the marrow spaces all over the body yet it may develop as a solitary lesion [11]

A solitary plasma cell myeloma of the skull apparently does occur Willis[12] thought they could be so classified if no Bence Jones proteinuria or no other lesion appeared in 1 year Vandenberg and Coley[13] reported 2 cases that were apparently cured with high voltage roentgenotherapy A needle biopsy helps to rule out other lesions, such as, metastatic cancer hemangioma, eosinophilic granuloma or early primary malignant bone tumors

In case of multiple myelomas involving the calvaria the myeloma tissue may destroy the diploic structures and fill the space. There also appears thinning of the tables in discrete punched out areas In these areas the tables are seldom penetrated Usually when the skull is involved by the process, many of these areas, varying in size from a pin head to several centimeters in diameter will be found by roentgenographic study, depending upon the stage of the disease In the far advanced cases, many bones will be involved and the skull defects may be larger

Pain is the most common symptom Weakness and anemia are quite common. A leukocytosis with plasma cells predominating may occur but is rare, and more apt to be found in patients under 50 than those above that age Bence Jones proteinuria, sometimes in the course of the disease, can be found in over 50% of cases An increase of serum globulin occurs in some cases Hyperproteinemia[14] with abnormal beta and gamma globulins may suggest the diagnosis in some cases The use of paper electrophoresis to pick up the presence of abnormal globulins[15] provides a very valuable diagnostic aid Serum phosphatase is not

altered A bone marrow study is one of the best means of establishing a diagnosis

Therapeutically roentgenotherapy will often relieve pain and may prolong life The course of this disease can seldom be predicted making it difficult to tell just how much therapeutic agents benefit the patient Radioactive phosphorus stilbamidine and pentamidine have been used, with varying degrees of success Surgery is indicated for relieving focal areas of pressure, as in cord compression Otherwise it can be of little help Since the tumor tissue in the radiolucent punched-out areas seldom penetrates the inner table of the skull and there is no tendency to penetrate the dura surgery for focal cerebral compression is not necessary

CASE REPORT

C. H B., female age 75 years, was referred in 1950 because of pain in the lower cervical and midthoracic area She also complained of pain in the left arm and hand These symptoms were first noted about 6 months previously It was learned that she had lost considerable weight and strength. It was noted that she was rather pale with a tendency towards cachexia There was tenderness over C-7 Some hypesthesia was elicited over the ulnar distribution on the left There were 2 540 000 red blood cells, color index of 86 WBC was 5 700 with lymphocytes 46 and neutrophils 54 Hematocrit was 24 cc packed cells/100 cc blood Sedimentation rate was 30 in 10 minutes. There was a trace of albumin. Serum albumin was 3 35 serum globulin 5 53 and serum protein 8 88

The roentgenograms revealed almost complete destruction of the body of C-7 T-8 and T 12 and the first rib on the left. There was increased density in the left apex There were numerous punched-out areas in the skull particularly in the parietal regions. Bone marrow smears showed a predominance of plasma cells.

Roentgenotherapy was given at intervals as palliative measure. Also ethyl carbamate was given for tolerance and maintenance dosage Some temporary relief was obtained Blood transfusions for a while improved the tired feeling The cachexia gradually increased, and she expired about 6 months later

RETICULUM CELL SARCOMA

Reticulum cell sarcoma was first described in 1939 by Parker and Jackson[16] It occurs in preadult, as well as later years of life Coley[1] tabulated the findings and results in 23 cases In these, the ages ran from 8 to 63 years. It is interesting to note that in 12 of this group the disease had been eradicated in 5 treatment was in progress, and in 5 the patient had died of the disease The prognosis is much better than in Ewing's tumor with which it is so often confused Also it may be noted that in none of the above cases did the lesion occur in the calvaria.

Ivias and Dahlin[17] found 49 reticulum cell sarcomas in 2,000 cases of primary bone tumors There was 1 among these 49 cases that was located in the left parietal region They noted that the prognosis was better than most of the other primary bone tumors

Only recently Ullrich and Bucy[18] reported the third case of reticulum cell sarcoma of the skull to be recorded Through the kindness of the above authors parts of this case record are here reported We are also grateful to Charles C Thomas, publisher and the Editor of the *American Journal of Roentgenology, Radium Therapy and Nuclear Medicine* for permission to reproduce portions of this work

CASE HISTORY

The patient was a 53 year old white female, whom they first saw in August 1952 complaining of pains in her head for 8 months. There were tender lesions 8 x 9 cm. in the left frontal and 5 x 5 cm. in the left occipital areas Roentgenograms showed progressive bone destruction, which, in view of the history of and extensive treatment for pulmonary tuberculosis, was diagnosed tuberculous osteomyelitis of the skull

In August 1952 a left frontal craniectomy was performed A mass of granulation tissue 2 x 3 cm. was first removed and then the soft eroded bone was rongeured away until normal bone was encountered A defect in the skull 8 x 9 cm was produced They reported that the dura was not involved A like, but smaller lesion in the left occipital area was treated in the same manner 5 days later

The microscopic examination as reported by the authors, was as follows The cells were imperfectly subdivided by trabeculae

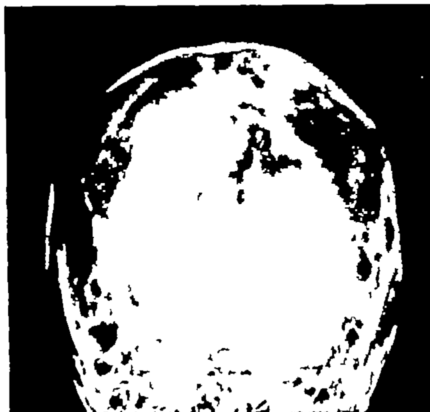


Fig 40a. Reticulum cell sarcoma of the calveria (from Ulrich and Bucy)

and small blood vessels, which were accompanied by a scanty collagenous stroma This loose stroma was reinforced by the characteristic network of fine reticulin fibers, which were well demonstrated by impregnation with silver The cells of the tumor ranged from ordinary marcophage like and fibrocyte like cells to large cells with obulated or multiple nuclei In some the nuclei were quite large Mitotic figures were numerous. They were found in all stages. Many of them were bizarre. The bone marrow had disappeared from the region of the tumor The few small dark cells present were lymphocytes and plasmocytes. Bone persisted focally especially near the margins, as eroded trabeculae or necrotic fragments. Occasionally bits of reactive new bone were seen The histopathologic diagnosis was typical highly malignant reticulum cell sarcoma.

This patient was studied at intervals for a period of 5 years. Roentgenotherapy was given postoperatively (3 500 r to 4 000 r

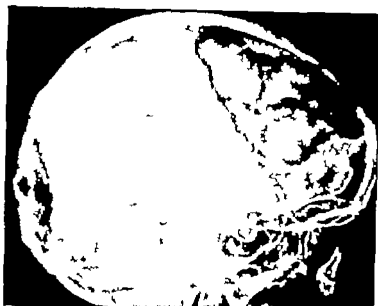


Fig 40d Same case, showing postoperative skull defect.



Fig 40c Same case, showing low power
Fig 40d. Same case, showing densely packed pleomorphic cells with numerous mitotic figures.

to each area) Repeated roentgenograms of the skull failed to reveal further destruction of bone, and the patient has remained well as far as the tumor was concerned

Since there is information available in the literature concerning only 4 cases of primary reticulum cell sarcoma of the skull

no definite conclusions may be made in regard to treatment. Two cases of Strange and DeLorimier[19] received only 1,000 r. and 1,800 r., respectively, and remained free of symptoms after many years. Bucy concluded that roentgenotherapy, after a biopsy diagnosis, is probably the best treatment. The surgical removal of the lesion, followed by a plastic cranioplasty, seems simpler and obviates the usage of large dosage of roentgenrays over the brain.

HEMANGIO-ENDOTHELIOMA (MALIGNANT)

There is considerable controversy concerning the classification of this lesion as an entity. Some authorities have shown preference for another term, or the inclusion of this lesion within another group of tumors. It has been suggested[20] that only those tumors in which microscopic evidence of malignant tumor with vasoformation, should be included in this group.

This is a rare tumor. Pancoast, Prendergrass and Schaeffer[5] have mentioned the involvement of the skull, stating that a dilated vascular channel may be the only roentgenographic evidence until late in the disease. Although we have no microscopic evidence to verify the tumor forming blood vessels, clinically, in the case reported below, there were multiple enlarged vascular channels, bruits and eventually destruction of bone and pulsation. Even in spite of multiple ligation of the primary arteries to the head, blood supply remained ample, probably suggesting new blood channel formation.

CASE REPORT

R. P., Male, age 13 years, was referred in 1948 because of headache, progressively increasing during the past 2 months. It was generalized. It would awaken him frequently during the night. There were no signs of increased intracranial pressure and no focal signs. The positive physical findings were confined to the skull. The right side of the skull was larger than the left. Also, numerous enlarged vascular channels were noted in the scalp and in the skull on the right side. Some right occipital and suboccipital tenderness was elicited. Pneumoencephalograms disclosed a slightly enlarged right lateral ventricle, with the septum

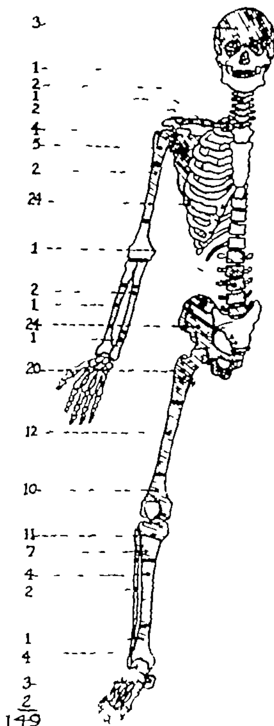


Fig 41 Location of primary tumor in 149 cases of endothelioma (from *Neoplasms of Bone* Courtesy Bradley L. Coley M.D., author and Paul B. Hoeber Inc publisher)

shifted to the left An angiogram (carotid) revealed a faint accumulation of contrast media in the right occipital area beneath a burr hole that had been previously placed for a ventriculogram. (Bleeding from the skull and dura was so brisk at that time that introduction of oxygen into the right ventricle had been abandoned) Section of bone taken from this area did not reveal any evidence of neoplasm

The headaches continued to be his chief complaint Tenderness and soreness in the right occipital area increased A large area of decreased density gradually became more prominent, and in June 1950 bone destruction was noted by palpation and roentgenograms. A bruit was audible and later a pulsation was palpable At that time a partial left homonomous hemianopsia was present

Compression of the right carotid artery stopped both the bruit and the pulsation The right common carotid and vertebral arteries were ligated in August 1950 Roentgenotherapy was given and symptoms abated almost completely The bruit ceased and the bulging at the skull defect receded (School, basketball and hunting were engaged in along with other normal activities of a boy of 15)

Early in 1952, prominence of scalp vessels was again noted Another area of decreased density in the skull was noted this time in the right parietal area A bruit could be heard over this region Compression of the left carotid artery relieved it. In July 1952 the left common carotid artery was ligated The bruit and pulsation subsided The bulging at the cranial defects receded No untoward effects from the occlusion of the carotid were noted He left the hospital in less than a week, feeling much improved

Up to this time, cerebral function remained unimpaired Roentgenograms of the chest did not reveal evidence of metastasis There were no symptoms directed to any other part of the body except the head and all complaints continued to be referrable to the cranial problem He continued to be an A student was active in basketball and also 4-H work to within a few months of his death

In September 1952 a little over 4 years after the first symptoms developed this patient succumbed to what seemed to be a respiratory failure. He was home at the time and unfortunately a postmortem examination was not secured

It seems most likely that primary vascular changes accounted

for the marked vascularity in the bone and dura in 1948, when a ventriculogram was attempted and for the later destruction of the skull in 2 different areas. A bruit was heard, a pulsation felt and large vascular channels noted in the scalp and by roentgenograms. There was a pooling of contrast media before the ligation of the carotid. It would seem that new vascular channels were responsible for progressive changes associated with bruit, pulsation and bone destruction. Then also, there was an expansion of the skull on the right side. An expansion of the shaft of long bones occurs quite commonly when involved by endothelioma.

The reader is referred to the recent work of Carter and Dickerson [21] who have furnished a good review of the literature and reported 1 more case.

CHLOROMA

Chloroma is a disease that originates from the lymphatic or myeloid cells of the bone marrow. Tumor infiltration may involve most any tissue of the body. The calvaria is affected in a high percentage of cases. It occurs in younger individuals, but at times may be found in later adult life. It is characterized by a tumor like growth that varies in size, depending upon the stage of the disease, but may become quite large, forming a large tumorous mass. The skull may be penetrated or it may even extend intracranially. The dura is seldom penetrated. The faint greenish color lends the tumor its name. Growth is rapid and death ensues in 1 to 2 years.

BIBLIOGRAPHY

1 Coley Bradley L. *Neoplasm of Bone* New York Hoeber 1949
2 Geschickter C. F Copeland M M Tumors of Bone (ed 2) *Am J Cancer* 1936 p 669
3 Cade, Sir S Osteogenic sarcoma, a study of 133 patients, *J Roy Coll Surg, Edinburgh, 1* 79 111 December 1955
4 Krantz, S Gay B. B Jr Primary chondrosarcoma of the occipital bone. *Am J Roentgenol* 69 598-604 April, 1953
5 Pancoast, H. K. Pendergrass, E. P Schaeffer J P *The Head and Neck in Roentgen Diagnosis* Springfield, Thomas, 1940 p 251
6 Geschickter C F So-called fibrosarcoma of bone bone involvement by sarcoma of neighboring soft parts. *Arch. Surg* 24 231 1932

7 Lichtenstein, I *Bone Tumors* St Louis, Mosby 1952, p 180
8 Dandy W E. *Dean Lewis Practice of Surgery* Hagerstown Prior 1932, Vol XII
9 Kirshbaum J D Fibrosarcoma of the skull in Paget s disease. *Arch Path* *36* 74 79 July 1943
10 Ewing J Diffuse endothelioma of bone *Proc N Y Path Soc* *21* 7 1921
11 Schwartz C. W Solitary myeloma of the frontal bone *Am J Roentgenol* *53* 573-574 June 1945
12. Willis, R. A Solitary plasmacytoma of bone *J Path & Bact* *53* 77 1941
13 Vandenberg H J Jr Coley B L. Primary tumors of the cranial bones. *Surg Gynec & Obst* *90* 602-612, May 1950
14 Wintrobe, M M Buell M V Hyperproteinemia associated with multiple myeloma with report of a case in which an extraordinary hyperproteinemia was associated with thrombosis of retinal veins and symptoms suggesting Raynaud s disease *Bull John Hopkins Hosp.*, *52*. 156 1933
15 Volpe, Robt Bruce-Robertson Alan, Fletcher Almon, Charles, W Bruce Essential cyro globulus, review of literature and report of a case treated with ACTH and cortisone *Am J Med* *20* 533 January June 1956
16 Parker F Jr Jackson H Jr Primary reticulum cell sarcoma of bone. *Surg Gynec & Obst* *68* 45 1939
17 Ivias, J C. Dahlin, D C. Reticulum cell sarcoma of bone. *J Bone & Joint Surg* *35 A* 835-842 October 1953
18 Ullrich, D P Bucy P *Am J Roentgenol Rad Therapy & Nuclear Med.*, *79* 4 April 1953
19 Strange, V M DeLorimier A A Reticulum cell sarcoma primary in skull report of 3 cases. *Am J Roentgenol Rad Therapy & Nuclear Med* *71* 40-50 1954
20 Hauser H Angiosarcoma of bone. *Am J Roentgenol* *42*. 656 1939
21 Carter J H Dickerson R. et al Angiosarcoma of bone A review of the literature and presentation of a case. *Ann. Surg* *144* 107 117 July 1956

ADDENDUM

This recent case of intradiploic traumatic cyst in an 8 weeks old infant, has just been seen and has been added because it is a classical representation of this type of lesion

M A M., 8 weeks of age first seen in June 1959 because of a lump on the right side of the head noticed immediately after birth It had not changed in size, and now was very firm yet for a short time after birth there was some fluid directly overlying the mass. Roentgenograms showed an area of increased density measuring 5 x 5 cm and extending outward from the normal contour of the skull approximately 1½ cm. The tangential view revealed a dense outer plate, while the remainder of this mass appeared cystic, with a few radiopaque areas interspersed The inner table was intact.

At operation the outer table was removed exposing a grayish/yellow soft fibrous mass which could be curetted away from the inner table. The outer table was removed completely the inner table was left intact The sections of this soft material revealed well-organized fibrous tissue, and scattered throughout this tissue were areas of hemosiderin pigment It was considered an organized intradiploic hematoma



Fig 1 Intradiploic traumatic cyst, lat. view

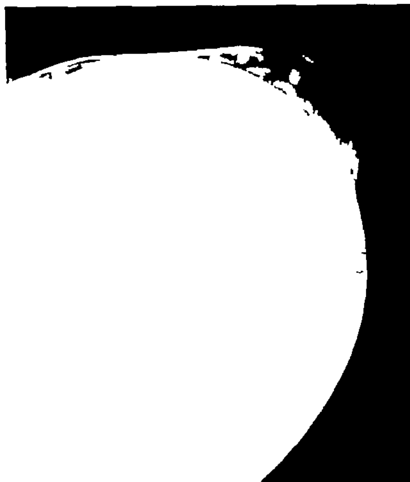


Fig. 2. Intradiploic traumatic cyst: tangential view

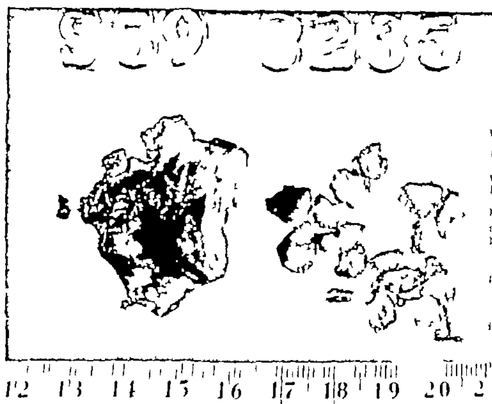


Fig 3 Same — L fibrous tissue mass removed from cystic cavity R bone removed—outer table.

INDEX

M

N

O

P

R

S